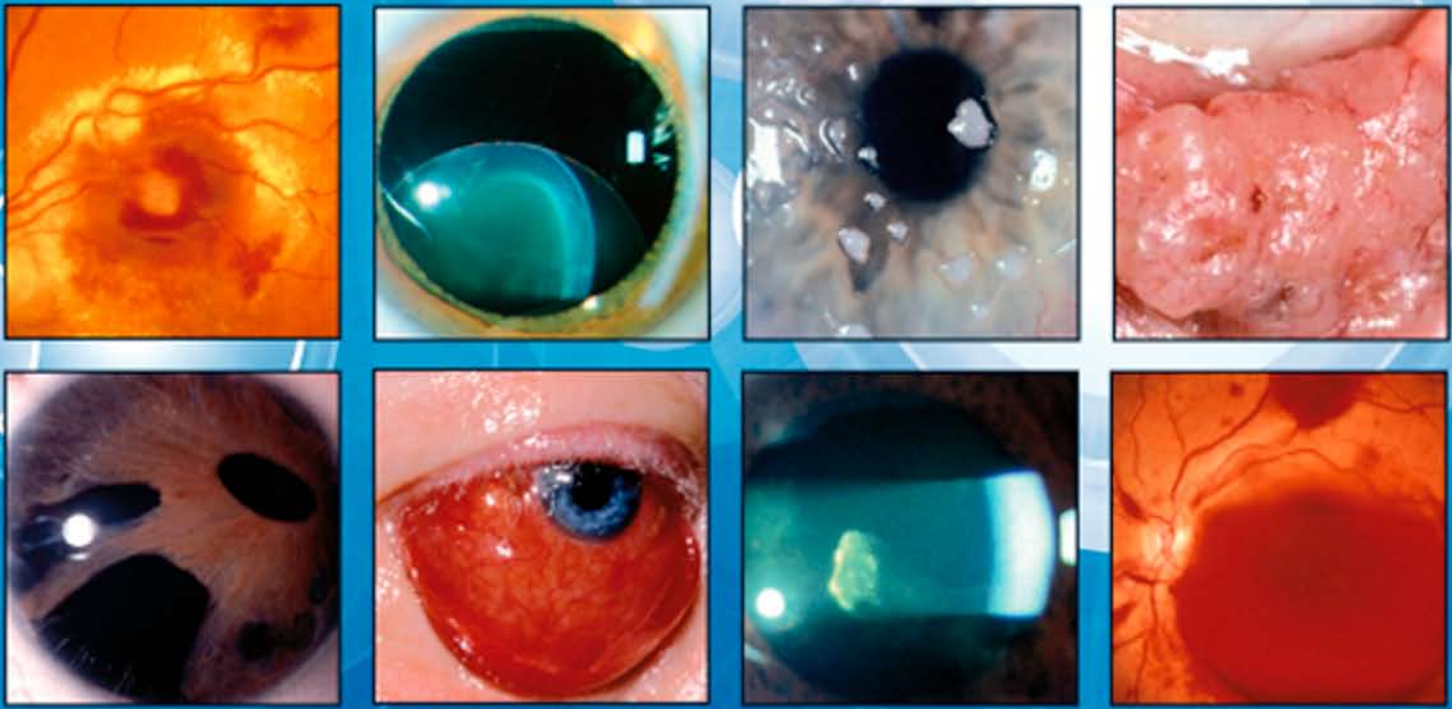
Jack J

Kanski

Signs in Ophthalmology

Causes and differential diagnosis



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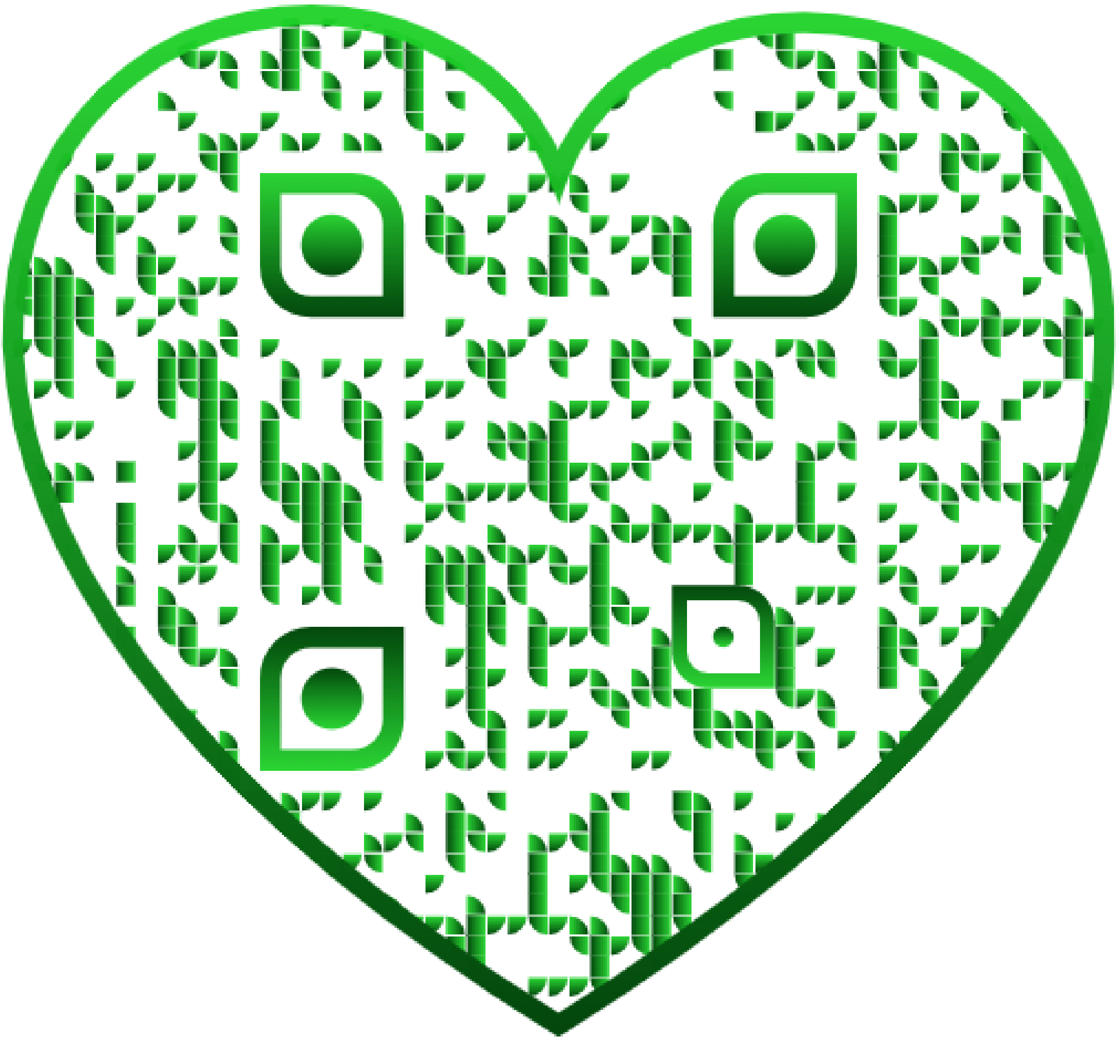

Signs in Ophthalmology
Causes and differential diagnosis

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Signs in Ophthalmology

Causes and differential diagnosis

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Contents

Preface *vi*
Acknowledgements *vii*

1 Eyelids 1
2 Orbit 39
3 Conjunctiva 61
4 Episclera and sclera 91
5 Cornea 101
6 Anterior chamber 141
7 Intraocular pressure and angle 151
8 Iris 163
9 Pupils 193
10 Lens 211
11 Vitreous 235
12 Optic nerve head 249
13 Fundus 275
14 Ocular motility 355
15 Systemic signs 383

Index *421*

Detailed chapter contents lists can be found at the beginning of each chapter.

Preface

Much of the diagnosis of ocular disease is based on the accurate observation and correct interpretation of clinical signs. The main purpose of this book is to acquaint the reader with the salient characteristics, causes and differential diagnosis of the many signs that may occur in ophthalmic disorders. The last chapter is devoted to systemic signs that may occur in diseases that have ophthalmic manifestations. Where appropriate, special investigations such as fluorescein angiography and MR are also included. The text is concise yet comprehensive and key colour-coded information is used to help the reader to navigate through each section. This book should be of particular interest to ophthalmologists in training and optometrists.

Jack J. Kanski
Windsor 2009

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Heyreh S. 12.27, 12.40. **Jager M.** 1.104, 3.70, 3.84, 5.57, 10.43, 10.49, 10.52, 10.53, 10.57.

Krachmer JH, Mannis MJ, Holland EJ. *Cornea,* Mosby 2005. 1.45, 1.114, 1.116, 3.28, 3.52, 3.87, 3.92, 5.23, 5.41, 5.50, 5.51, 5.75, 5.89, 5.94, 5.96, 5.97, 8.10. **Leys A.** 13.243. **Link T.** 12.51. **Lisch W.** 3.66.

MacKeen L. 1.34, 2.12, 2.28, 7.6, 7.12, 7.17, 7.24, 9.34. **Martinkova R.** 8.43.

McAllister J. 8.53. **Merin L.** 1.37, 5.90, 12.15, 13.35, 13.84, 13.124, 13.222, 13.254, 13.255, 15.26. **Messmer E.** 11.35. **Meyer D.** 1.128. **Milweski SA.** 13.226, 13.227, 13.268. **Mir A. *Atlas of Clinical Diagnosis*, Saunders 2003.** 3.37, 4.17, 5.112, 8.18, 8.95, 10.11, 13.97, 13.100, 15.38, 15.57, 15.64, 15.68, 15.69, 15.87, 15.93, 15.98, 15.106. **Moorfields Eye Hospital.** 13.98, 13.225, 13.236, 13.278. **Morse PH.** 12.49, 13.82, 15.13. **Nerad JA, Carter KD, Alford, MA. *Oculoplastic and Reconstructive Surgery*, *in Rapid Diagnosis in Ophthalmology*, Mosby, 2008.** 1.74, 1.82, 2.3, 2.37, 4.18.

Nischal KK. 1.127, 2.11, 2.19a, 2.21, 5.27, 5.93, 7.30, 8.73, 11.11, 12.52, 13.253, 15.16. **Noble B.** 8.17. **Parluekar M.** 1.76, 5.18, 14.22. **Pavésio CE.** 8.5, 11.5, 11.28, 13.102, 13.130, 13.164, 15.25. **Pearson A.** 1.2, 1.6, 1.79, 1.104, 1.108, 1.125, 1.126, 2.35. **Puri K.** 5.62. **Raik N.** 15.63. **Raina UK.** 1.47, 5.24, 7.29, 8.25, 8.98, 11.25. **Ridgway A.** 5.102. **Rogers N.** 1.85, 1.111, 5.14, 5.23, 5.79, 6.2, 7.34, 8.54, 9.16, 9.18, 9.32, 9.33, 10.2, 10.32, 15.14.

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Eyelids 1

DISORDERS OF LASHES 3
Madarosis 3
Poliosis 4
Misdirection 4
Trichiasis 4
Congenital distichiasis 5
Acquired distichiasis (metaplastic lashes) 5
Eyelash ptosis 5
Epiblepharon 6
Trichomegaly 6

INFLAMMATORY LID MARGIN DISEASE 7
Staphylococcal blepharitis 7
Seborrhoeic blepharitis 7
Posterior blepharitis 7
Childhood blepharokeratoconjunctivitis 8
Angular blepharitis 8
Phthiriasis palpebrarum 8

SUPERFICIAL INFLAMMATION 9
Allergic 9
Contact dermatitis 9
Atopic dermatitis 9
Infectious 10
Herpes zoster ophthalmicus 10
Impetigo 10
Erysipelas 10
Necrotising fasciitis 10

DIFFUSE EYELID SWELLING 11
Inflammatory causes 11
Associated with conjunctivitis 11
Associated with dacryoadenitis 11
Associated with dacryocystitis 11
Inflammatory orbital disease 12
Angioedema 12
Local non-inflammatory causes 12
Orbital fat herniation 12
Blunt trauma 12
Blepharochalasis 13
Direct carotid–cavernous fistula 13
Rapidly growing tumours 13
Systemic causes 13
Myxoedema 13
Primary amyloidosis 14
Renal failure 14
Adult-onset periocular xanthogranuloma 14
Rosai–Dorfman disease 14

DISCOLORATION OF EYELIDS 15
Oculodermal melanocytosis (naevus of Ota) 15
Sturge–Weber syndrome 15
Lentigo maligna (Hutchinson freckle) 15
Congenital naevus 16
Dermatomyositis 16
Vitiligo 16
Minocycline 16

LID RETRACTION 17
Thyroid eye disease 17
Myasthenia gravis 17
Third nerve misdirection 17
Marcus Gunn jaw-winking syndrome 17
Collier sign of the midbrain (Parinaud syndrome) 18
Miscellaneous causes 18

PTOSIS 19
Neurogenic adult ptosis 19
Third nerve palsy 19
Horner syndrome (oculosympathetic palsy) 20
Third nerve misdirection 20
Myogenic adult ptosis 20
Myasthenia gravis 20
Myotonic dystrophy 21
Ocular myopathy 21
Other types of adult ptosis 22
Aponeurotic 22
Mechanical 22

NEUROGENIC CHILDHOOD PTOSIS 23
Marcus Gunn syndrome 23
Horner syndrome (oculosympathetic palsy) 23
Third nerve palsy 23
Bassen–Kornzweig syndrome 23
Myogenic childhood ptosis 24
Simple congenital ptosis 24
Blepharophimosis syndrome 24
Congenital fibrosis syndrome 24
Myasthenia gravis 25
Causes of pseudo-ptosis 25
Brow ptosis 25
Dermatochalasis 25
Contralateral lid retraction 25
Ipsilateral hypotropia 26
Lack of lid support by the globe 26
Lash ptosis 26

MULTIPLE PAPULES AND VESICLES 26
Herpes simplex 26
Molluscum contagiosum 27
Milia 27
Syringomas 27
Comedones 27
Dermatosis papulosa nigra 28
Xanthelasma 28

CYSTIC LESIONS 28
Apocrine sweat gland hidrocystoma (cyst of Moll) 28
Eccrine sweat gland hidrocystoma 28
Cyst of Zeis 29
Pilar ('sebaceous') cyst 29
Epidermal inclusion cyst 29
Superficial dermoid cyst 29

PAPILLOMATOUS LESIONS 30
Squamous cell papilloma 30
Basal cell papilloma (seborrhoeic keratosis, senile verruca) 30
Inverted follicular keratosis 30

NODULAR LESIONS 31
Benign 31
Chalazion (meibomian cyst) 31
External hordeolum (stye) 31
Capillary haemangioma 31
Acquired naevus 32
Actinic (solar, senile) keratosis 32
Keratoacanthoma 32
Neurofibroma 32
Pilomatricoma (pilomatrixoma) 33
Trichilemmoma 33
Cutaneous horn 33

Malignant **33**
- *Nodular basal cell carcinoma* 33
- *Nodular squamous cell carcinoma* 34
- *Nodular meibomian gland carcinoma* 34
- *Kaposi sarcoma* 34
- *Nodular melanoma* 34
- *Merkel cell (neuroendocrine) carcinoma* 34

ULCERATING TUMOURS **35**
- *Ulcerating basal cell carcinoma (rodent ulcer)* 35
- *Ulcerating squamous cell carcinoma* 35
- *Ulcerating gland of Zeis carcinoma* 35

PLAQUE-LIKE LESIONS **36**
- *Bowen disease (intraepidermal carcinoma in situ)* 36
- *Sclerosing basal cell carcinoma* 36
- *Spreading meibomian gland carcinoma* 37
- *Superficial spreading melanoma* 37

MAJOR CONGENITAL ANOMALIES **37**
- *Ablepharon* 37
- *Microblepharon* 37
- *Coloboma* 38
- *Congenital lid inversion* 38
- *Cryptophthalmos* 38

DISORDERS OF LASHES

Madarosis

Signs

- Decrease in number or complete loss of lashes.

Causes

- See Table 1.1.

Table 1.1 **Causes of madarosis**

1. **Local**
    - Chronic anterior lid margin disease (**Fig. 1.1**)
    - Infiltrative lid tumours may cause localised madarosis (**Fig. 1.2**)
    - Burns (**Fig. 1.3**)
2. **Following removal**
    - Iatrogenic for treatment of trichiasis or distichiasis (**Fig. 1.4**)
    - Cryotherapy or radiotherapy of lid tumours
    - Trichotillomania which is a psychiatric disorder of habitual hair removal
3. **Generalised skin disease**
    - Generalised alopecia
    - Psoriasis
    - Atopic dermatitis (**Fig. 1.5**)
    - Ichthyosis (**Fig. 1.6**)
4. **Systemic disease**
    - Myxoedema
    - Systemic lupus erythematosus
    - Syphilis
    - Lepromatous leprosy

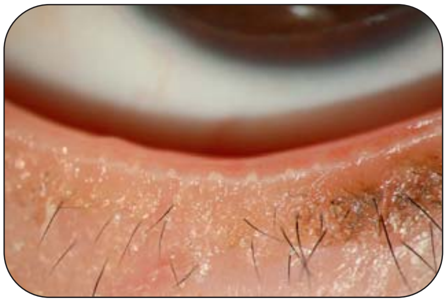
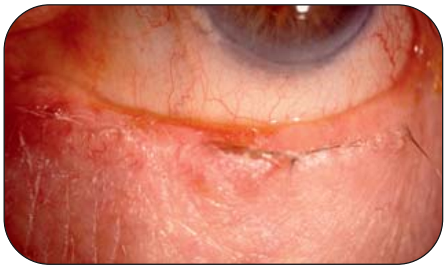


Fig. 1.1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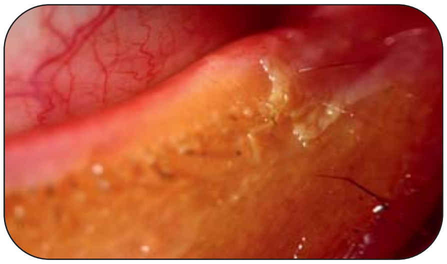
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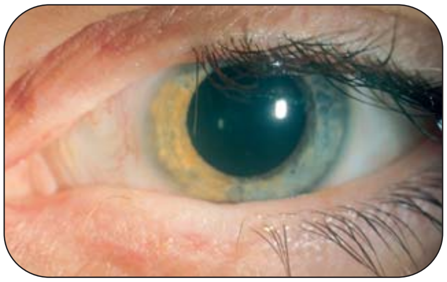
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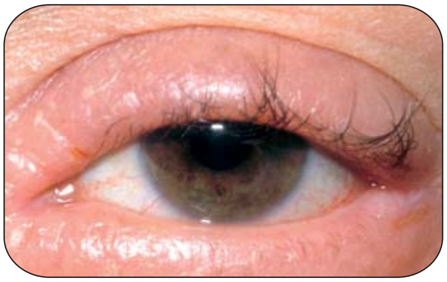
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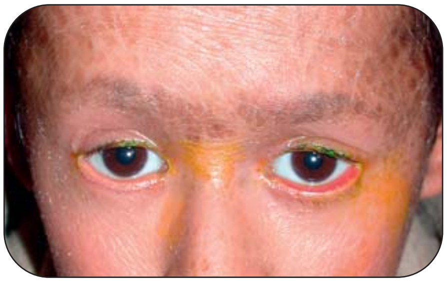


Fig. 1.6

Poliosis

Signs

- Localised patch of grey or white hair due to lack of pigment in the epidermis (**Fig. 1.7**).

Causes

- See Table 1.2.

Table 1.2 **Causes of poliosis**

1. **Idiopathic**
2. **Local conditions**
   - Chronic anterior blepharitis
   - Sympathetic ophthalmitis
   - Lid margin tumours
   - Herpes zoster ophthalmicus
   - Alopecia areata
3. **Systemic conditions**
   - Vogt–Koyanagi–Harada syndrome in which it is associated with vitiligo (**Fig. 1.8**)
   - Waardenburg syndrome
   - Marfan syndrome
   - Tuberous sclerosis

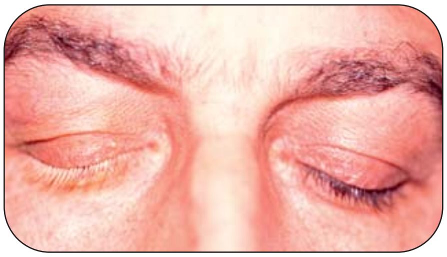


Fig. 1.7

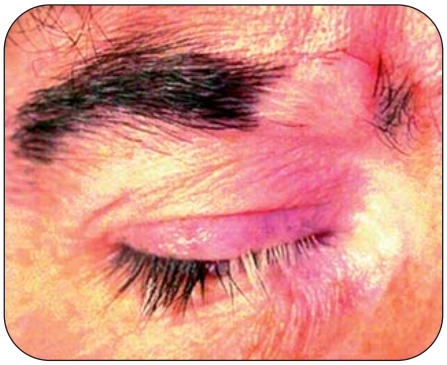


Fig. 1.8

Misdirection

Trichiasis

Signs

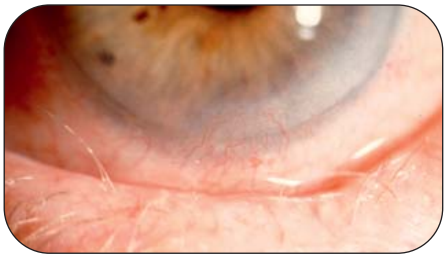
- Inward turning of previously normal lashes (**Fig. 1.9**).

Causes

- Idiopathic.
- Lid margin scarring (e.g. chronic blepharitis, herpes zoster ophthalmicus and trachoma).

Differential diagnosis

- Inward turning of lower lid lashes associated with entropion (pseudo-trichiasis – **Fig. 1.10**).



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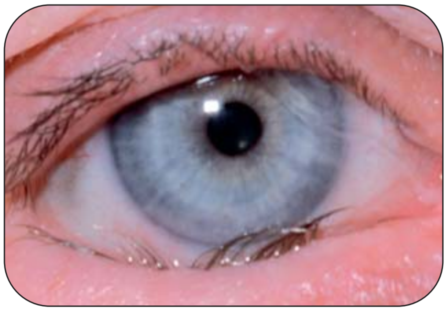

Fig. 1.10

Congenital distichiasis

Definition

- A rare, bilateral, hereditary condition.

Signs

- Partial or complete second row of lashes originating from or slightly behind the meibomian gland orifices (**Fig. 1.11**).

Look for

- Lymphoedema.

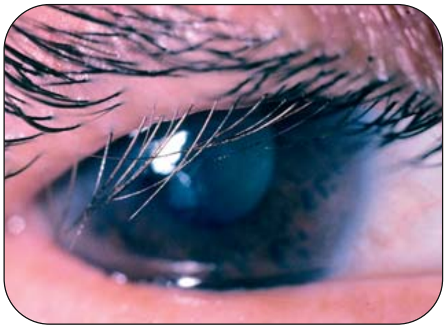
Fig. 1.11

Acquired distichiasis (metaplastic lashes)

Definition

- An uncommon unilateral or bilateral condition caused by metaplasia and dedifferentiation of meibomian glands to become hair follicles in late cicatrising conjunctivitis (e.g. ocular cicatricial pemphigoid).

Signs

- Crops of stunted lashes originating from meibomian gland orifices (**Fig. 1.12**).

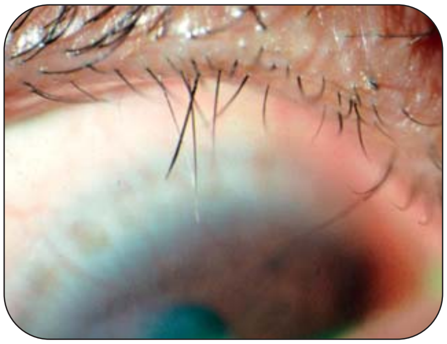
Fig. 1.12

Eyelash ptosis

Signs

- Downward drooping of upper-lid lashes (**Fig. 1.13**).

Causes

- Idiopathic.
- Floppy eyelid syndrome.
- Long-standing facial palsy.

Differential diagnosis

- True ptosis.
- Upper lid entropion.

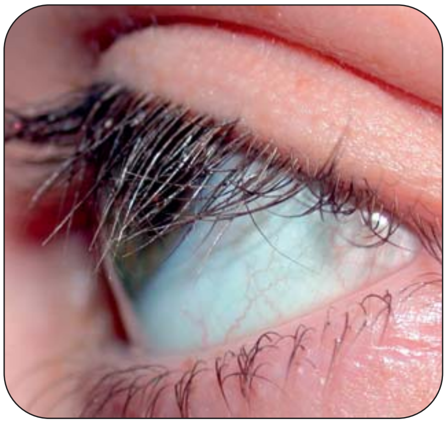
Fig. 1.13

Epiblepharon

Definition

- A bilateral congenital condition that is common in Orientals.

Signs

- Extra fold of skin stretches across the anterior lid margin and the lashes are directed vertically, especially medially (**Fig. 1.14**).
- When the fold of skin is pulled down the lashes return temporarily to their normal position.

Differential diagnosis

- Congenital entropion is characterised by in-turning of the entire lid and lashes with absence of the lower lid crease (**Fig. 1.15**); when downward pressure is applied to the lid the entire lid becomes pulled away from the globe.

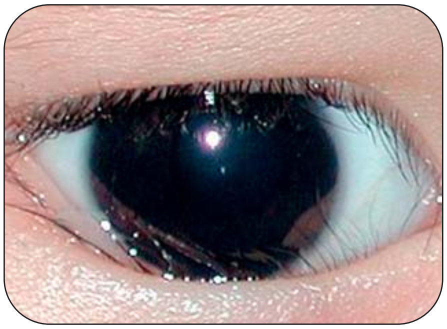

Fig. 1.14

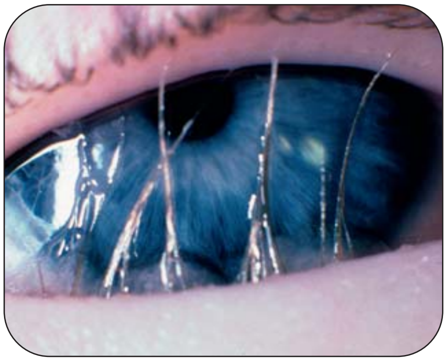

Fig. 1.15

Trichomegaly

Signs

- Excessive eyelash growth (**Fig. 1.16**).

Causes

- See Table 1.3.

Table 1.3 **Causes of trichomegaly**

1. **Congenital**
   - Oliver–McFarlane syndrome – RP, dwarfism and mental handicap
   - Cornelia de Lange syndrome – synophrys, low hairline, and developmental and musculoskeletal anomalies
   - Goldstein–Hutt syndrome – cataract and hereditary spherocytosis
   - Hermansky–Pudlak syndrome – albinism and bleeding diathesis
   - Oculocutaneous albinism type 1
2. **Acquired**
   - Drug-induced – phenytoin, ciclosporin and topical prostaglandin analogues (**Fig. 1.17**)
   - Malnutrition
   - AIDS
   - Porphyria
   - Familial

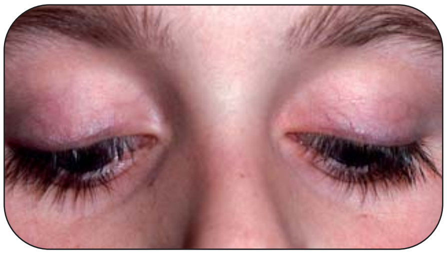

Fig. 1.16

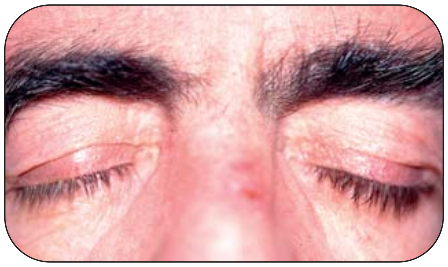

Fig. 1.17

INFLAMMATORY LID MARGIN DISEASE

Staphylococcal blepharitis

Signs

- *Lashes* – soft scales around lash roots (**Fig. 1.18**), madarosis, poliosis and trichiasis.
- *Anterior lid margin* – ulceration, notching and microabscesses.
- *Cysts* – acute external hordeolum (stye – see Fig. 1.102).
- *Tear film* – dry.
- *Conjunctiva* – papillae and phlyctens.
- *Cornea* – punctate erosions and marginal infiltrates.
- *Associated dermatitis* – atopic (see Fig. 15.1).

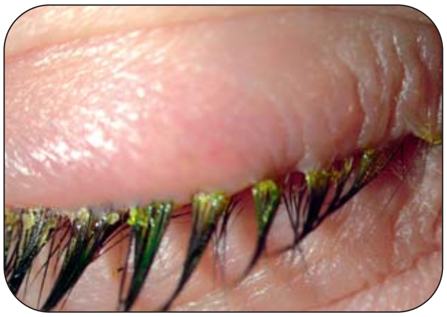

Fig. 1.18

Seborrhoeic blepharitis

Signs

- *Lashes* – soft greasy scales in between the lash roots and oily lashes stuck together (**Fig. 1.19**).
- *Anterior lid margin* – shiny.
- *Tear film* – dry.
- *Conjunctiva* – unremarkable.
- *Cornea* – punctate erosions and peripheral infiltrates.
- *Associated dermatitis* – seborrhoeic.

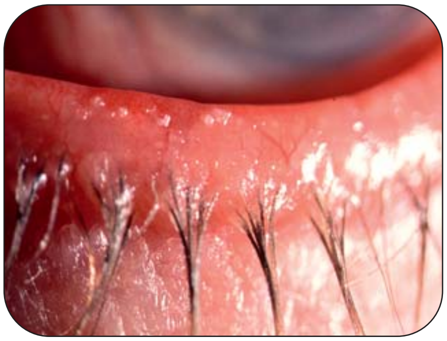

Fig. 1.19

Posterior blepharitis

Signs

- *Lashes* – unremarkable.
- *Posterior lid margin* – notching, oily capping or occlusion of meibomian gland orifices (**Fig. 1.20**), expressed meibomian secretions may be turbid and toothpaste-like.
- *Cysts* – meibomian (see Fig. 1.100).
- *Tear film* – dry and frothy.
- *Conjunctiva* – unremarkable.
- *Cornea* – punctate erosions and infiltrates.
- *Associated dermatitis* – acne rosacea (see Fig. 15.2).

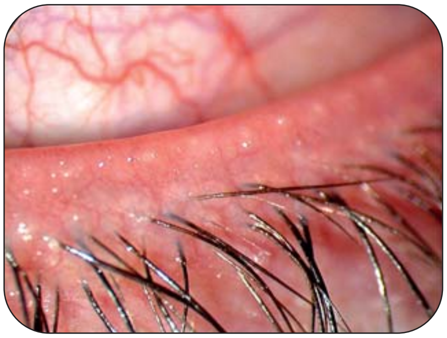

Fig. 1.20

Table 1.4 **Characteristics of chronic blepharitis**

		Anterior blepharitis		Posterior blepharitis
	Sign	**Staphylococcal**	**Seborrhoeic**	
Lashes	Deposit	Hard	Soft	
	Loss	++	+	
	Distorted or trichiasis	++	+	
Lid margin	Ulceration	+		
	Notching	+		++
Cyst	Acute hordeolum	++		
	Meibomian			++
Conjunctiva	Phlycten	+		
Tear film	Foaming			++
	Dry eye	+	+	++
Cornea	Punctate erosions	+	+	++
	Vascularisation	+	+	++
	Infiltrates	+	+	++
Associated dermatitis		Atopic	Seborrhoeic	Acne rosacea

Childhood blepharokeratoconjunctivitis

Signs

- *Lashes* – crusty.
- *Lid margin* – chronic anterior and posterior blepharitis.
- *Cysts* – styes and meibomian, often multiple (**Fig. 1.21**).
- *Tear film* – dry.
- *Conjunctiva* – phlyctens, and follicular or papillary hypertrophy.
- *Cornea* – punctate erosions, axial subepithelial haze, and peripheral infiltrates and vascularisation.
- *Associated dermatitis* – absent.

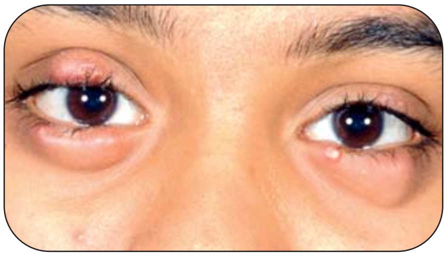

Fig. 1.21

Angular blepharitis

Signs

- *Lashes* – unremarkable.
- *Lids* – erythema, scaling and fissuring of skin at one or both canthi (**Fig. 1.22**).
- *Cysts* – absent.
- *Tear film* – unremarkable.
- *Conjunctiva* – follicular conjunctivitis.
- *Cornea* – marginal infiltrates and phlyctens.
- *Associated dermatitis* – atopic.

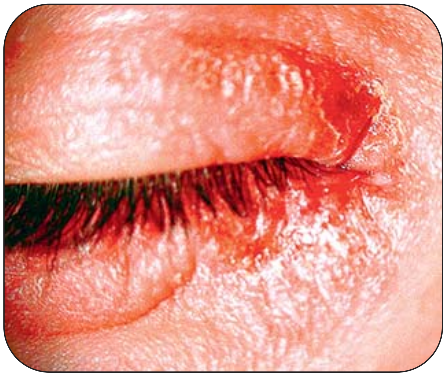

Fig. 1.22

Phthiriasis palpebrarum

Definition

- Infestation of the lashes with the crab louse (*Phthirus pubis*).

Signs

- *Lashes* – adherent lice and ova (**Fig. 1.23**).
- *Lids* – erythematous.

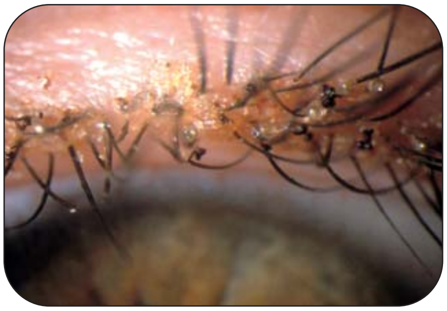
Fig. 1.23

SUPERFICIAL INFLAMMATION

Allergic

Contact dermatitis

Definition

- A common, unilateral or bilateral condition, frequently caused by sensitivity to topical medication.

Signs

- Initially there is erythema and oedema associated with ipsilateral conjunctivitis (**Fig. 1.24**).
- Thickening and crusting develops in long-standing cases (**Fig. 1.25**).

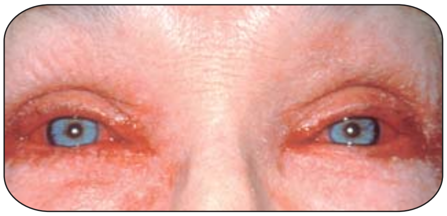
Fig. 1.24

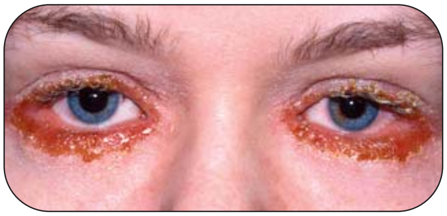
Fig. 1.25

Atopic dermatitis

Definition

- An uncommon, bilateral condition which may occur in patients with more generalised skin involvement.

Signs

- Erythema, thickening, vertical fissuring, often associated with madarosis and staphylococcal blepharitis (**Fig. 1.26**).

Look for

- See Table 1.5.

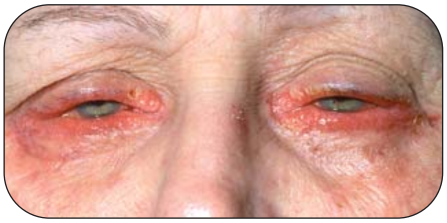
Fig. 1.26

Table 1.5 **Ocular associations of atopic dermatitis**

- Chronic staphylococcal blepharitis
- Angular blepharitis
- Vernal conjunctivitis in children
- Atopic keratoconjunctivitis in adults
- Keratoconus
- Anterior shield-like cataract
- Retinal detachment

Infectious

Herpes zoster ophthalmicus

Definition

- A common unilateral infection with varicella-zoster virus that typically affects the elderly.

Signs

- Painful maculopapular rash involving the first division of the trigeminal nerve followed by vesicles, pustules and crusting ulceration (**Fig. 1.27**).

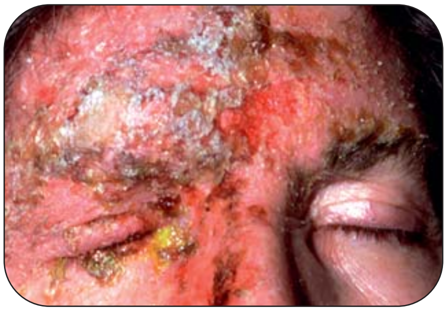

Fig. 1.27

Look for

- Hutchinson sign (involvement of the side of the nose).
- Anterior uveitis.
- Keratitis.
- Scleritis.
- Neurological complications.
- AIDS in young patients.

Impetigo

Definition

- A bilateral infection with staphylococci or beta-haemolytic streptococci that typically affects children.

Signs

- Small vesicles and bullae which on rupturing produce crusts composed of golden-yellow crystals (**Fig. 1.28**).

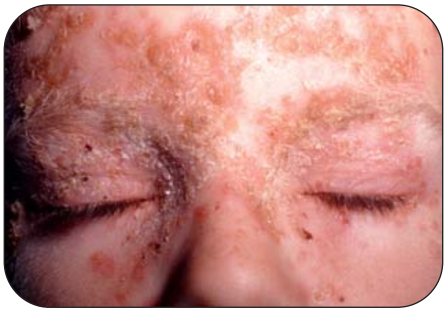

Fig. 1.28

Erysipelas

Definition

- A unilateral subcutaneous cellulitis caused by entry of beta-haemolytic streptococci at a site of minor skin trauma.

Signs

- Well-defined, erythematous, tender subcutaneous plaque, often with a butterfly configuration (**Fig. 1.29**).

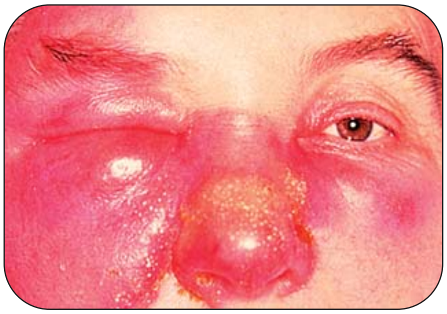

Fig. 1.29

Necrotising fasciitis

Definition

- A rare bilateral but life-threatening necrosis of subcutaneous soft tissues caused by *S. pyogenes* or *S. aureus.*

Signs

- Black discoloration due to gangrene secondary to underlying thrombosis (**Fig. 1.30**).

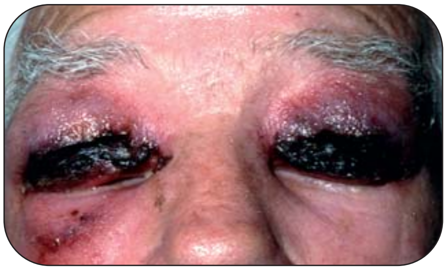

Fig. 1.30

DIFFUSE EYELID SWELLING

Inflammatory causes

Associated with conjunctivitis

Signs

- Adenoviral infection in particular (**Fig. 1.31**).

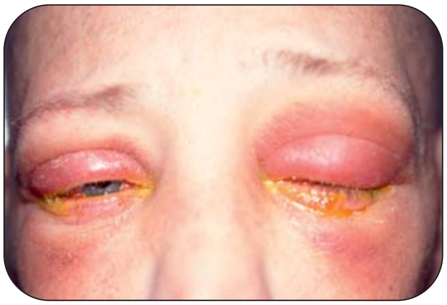

Fig. 1.31

Associated with dacryoadenitis

Signs

- Unilateral tender erythema and oedema involving the upper outer part of the lid causing a characteristic S-shaped deformity (**Fig. 1.32**).
- Injection of the lacrimal gland and conjunctiva may occur (**Fig. 1.33**).

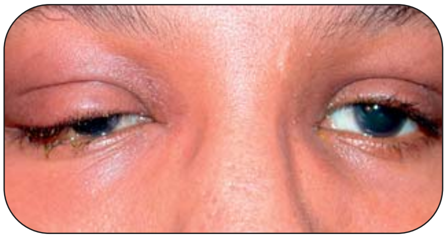

Fig. 1.32

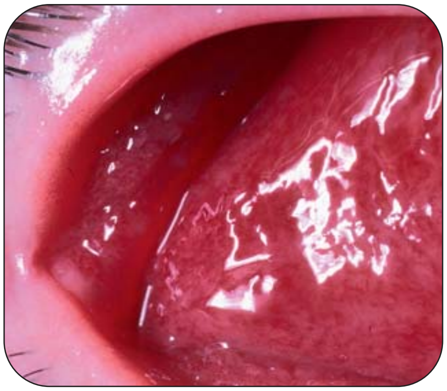

Fig. 1.33

Associated with dacryocystitis

Signs

- Very tender erythema and oedema at the inner canthus with spread to the lids (**Fig. 1.34**).

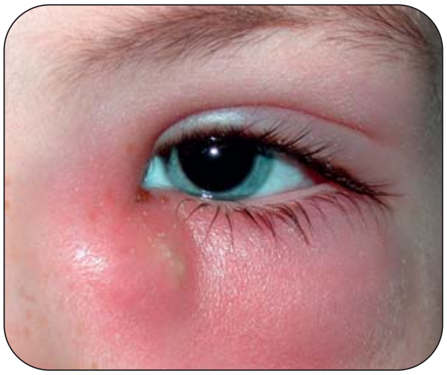

Fig. 1.34

Inflammatory orbital disease

Signs

- Unilateral or bilateral tender erythema and oedema, and mechanical ptosis (**Fig. 1.35**).

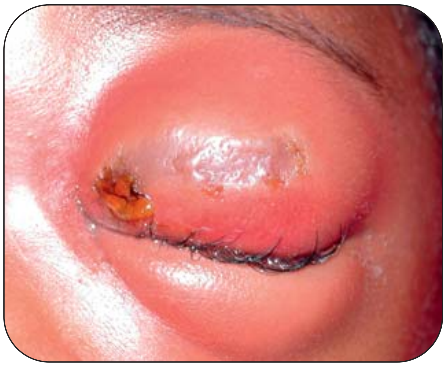

Fig. 1.35

Angioedema

Signs

- Transient painless pitting oedema (**Fig. 1.36**) that may be associated with oedema of the tongue or lips.

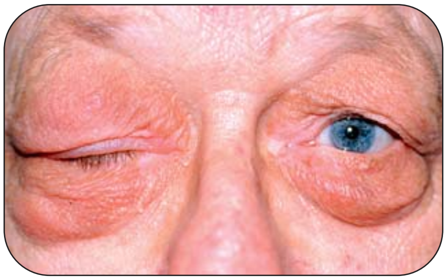

Fig. 1.36

Local non-inflammatory causes

Orbital fat herniation

Definition

- A common, usually bilateral, age-related condition.

Signs

- Asymmetrical periorbital swelling (**Fig. 1.37**).

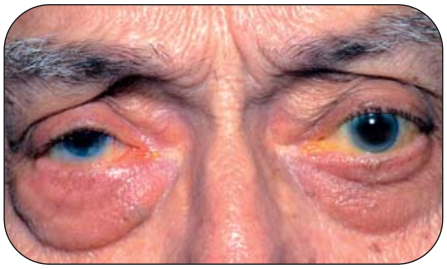

Fig. 1.37

Blunt trauma

Signs

- Painful oedema and ecchymosis (**Fig. 1.38**).
- Subcutaneous emphysema caused by medial orbital floor blow-out fracture (**Fig. 1.39**).

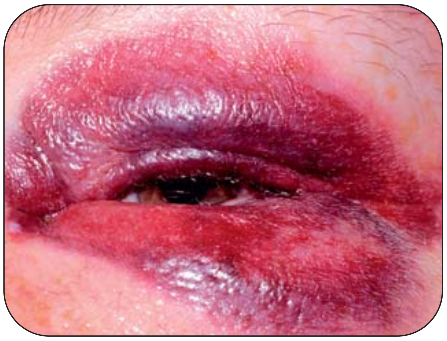

Fig. 1.38

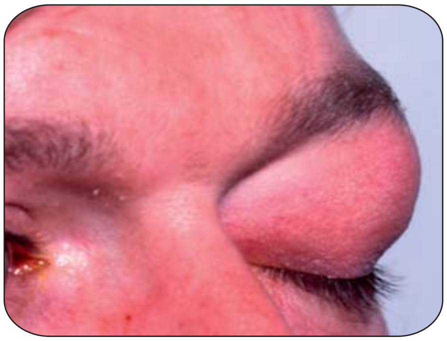

Fig. 1.39

Blepharochalasis

Definition

- An uncommon, usually bilateral condition which typically affects young individuals characterised by recurrent attacks of non-pitting oedema of the upper and occasionally the lower lid.

Direct carotid–cavernous fistula

Definition

- A high-flow shunt in which carotid artery blood passes directly into the cavernous sinus.

Signs

- Severe lid swelling and haemorrhagic chemosis (**Fig. 1.40**).

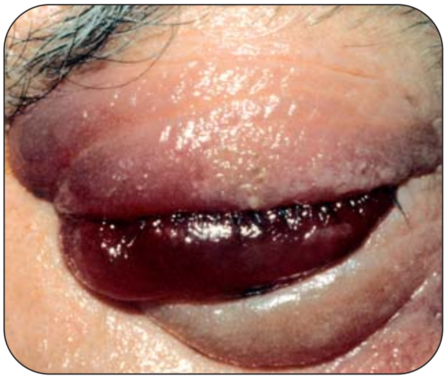

Fig. 1.40

Look for

- Pulsatile proptosis associated with a bruit.
- Anterior segment ischaemia.
- Raised intraocular pressure.
- Ocular motor nerve palsies.

Rapidly growing tumours

Signs

- In rhabdomyosarcoma the swollen lid is frequently red but not warm (**Fig. 1.41**).

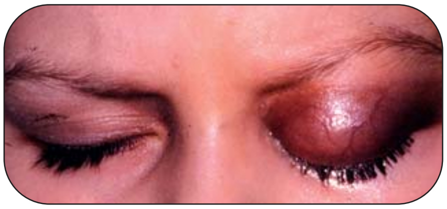

Fig. 1.41

Systemic causes

Myxoedema

Signs

- Swelling typically involves the lower lids (**Fig. 1.42**).

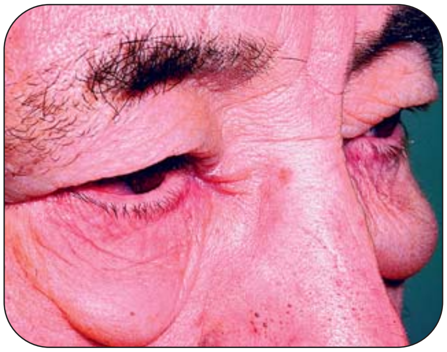

Fig. 1.42

Look for hypercholesterolaemia

- Xanthelasma.
- Arcus senilis.

Primary amyloidosis

Definition

- A rare condition in which there is accumulation of insoluble fibrillary proteins (amyloid) in various organs and tissues.

Signs

- Bilateral eyelid infiltration (**Fig. 1.43**).

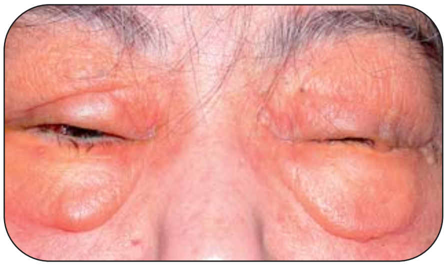
Fig. 1.43

Look for

- Involvement of other ocular structures such as the cornea, lacrimal gland, conjunctiva and orbit.

Renal failure

Signs

- Asymmetrical oedema mainly of the upper eyelids (**Fig. 1.44**).

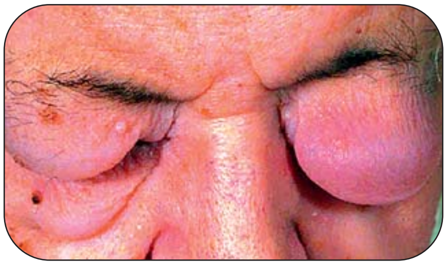
Fig. 1.44

Look for

- Ankle oedema and ascites.

Adult-onset periocular xanthogranuloma

Definition

- A very rare histiocytic granulomatous disease associated with asthma.

Signs

- Bilateral diffuse bilateral eyelid infiltration (**Fig. 1.45**).

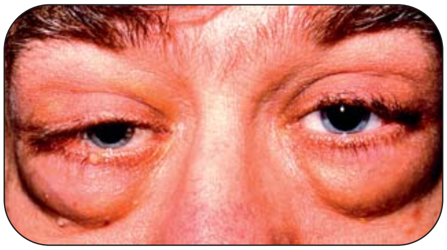
Fig. 1.45

Look for

- Subconjunctival infiltrates.

Rosai–Dorfman disease

Definition

- A rare disease usually seen in young individuals characterised by lymphadenopathy and constitutional features.

Signs

- Bilateral diffuse eyelid masses (**Fig. 1.46**).

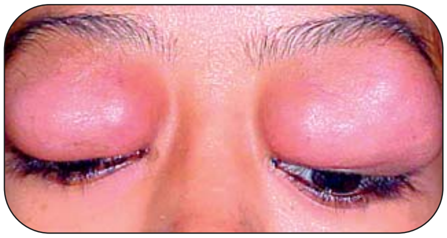
Fig. 1.46

DISCOLORATION OF EYELIDS

Oculodermal melanocytosis (naevus of Ota)

Definition

- An uncommon congenital usually unilateral condition.

Signs

- Grey-blue discoloration of the skin involving the distribution of one or more branches of the trigeminal nerve (**Fig. 1.47**).

Look for

- Conjunctival subepithelial melanosis (see Fig. 3.66).
- Iris hyperchromia (see Fig. 8.97) and mammillations (see Fig. 8.42).
- Hyperpigmentation of the trabecular meshwork (see Fig. 7.6) and fundus (see Fig. 7.7).
- Glaucoma.
- Increased risk of uveal melanoma.

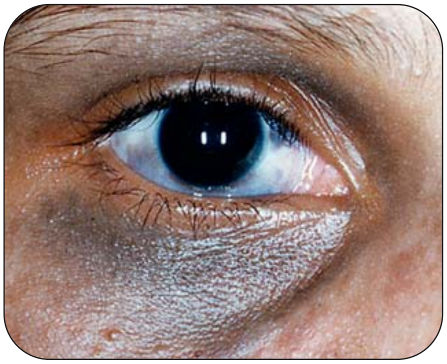

Fig. 1.47

Sturge–Weber syndrome

Definition

- A sporadic phacomatosis characterised by a port-wine stain and ipsilateral leptomeningeal haemangioma.

Signs

- Facial naevus flammeus (port-wine stain) involving the distribution of one or more branches of the trigeminal nerve (**Fig. 1.48**).

Look for

- Conjunctival telangiectasia (see Fig. 3.58).
- Iris hyperchromia.
- Glaucoma.
- Diffuse choroidal haemangioma.

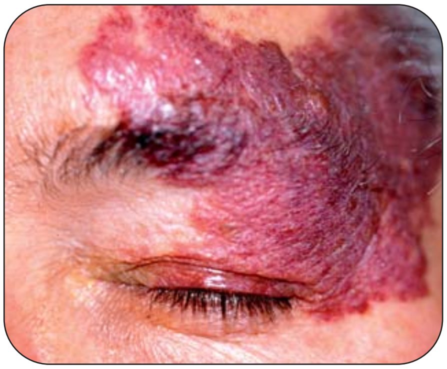

Fig. 1.48

Lentigo maligna (Hutchinson freckle)

Definition

- The pre-invasive stage of melanoma that typically affects elderly patients.

Signs

- Slowly expanding pigmented macule (**Fig. 1.49**).
- Nodular thickening is indicative of transformation into a frank melanoma.

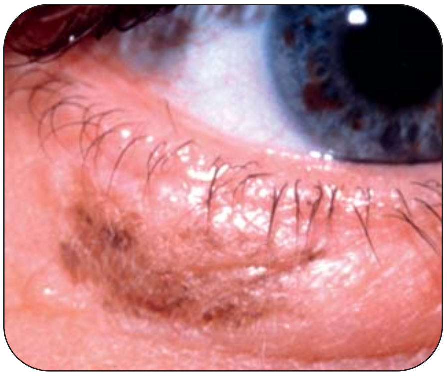

Fig. 1.49

Congenital naevus

Signs

- Usually fairly large and may occasionally involve both the upper and lower lids (split or kissing naevus – **Fig. 1.50**).

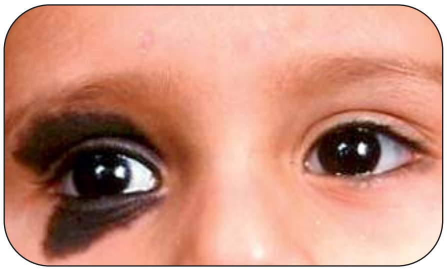

Fig. 1.50

Dermatomyositis

Definition

- An autoimmune disease characterised by involvement of skeletal muscle and skin.

Signs

- Heliotrope discoloration of the eyelids that may be associated with ulceration (**Fig. 1.51**).

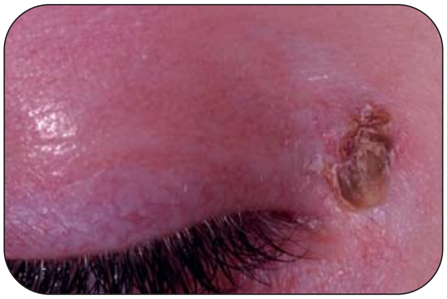

Fig. 1.51

Look for

- KCS.
- Scleritis.
- Retinopathy.

Vitiligo

Signs

- Well-defined symmetrical patches of progressive cutaneous depigmentation that may be associated with poliosis (see Fig. 1.8).

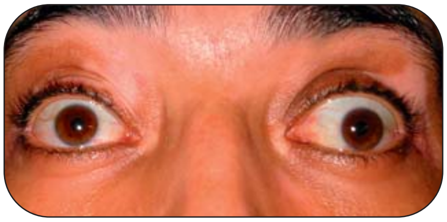

Fig. 1.52

Causes

- Idiopathic.
- Myxoedema.
- Thyrotoxicosis (**Fig. 1.52**).
- Pernicious anaemia.
- Autoimmune Addison disease.
- Vogt–Koyanagi–Harada syndrome.
- Sympathetic ophthalmitis.

Minocycline

Minocycline is a tetracycline that may occasionally cause cutaneous pigmentation (**Fig. 1.53**).

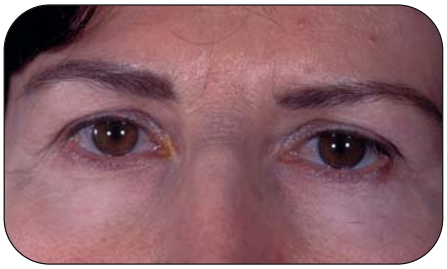

Fig. 1.53

LID RETRACTION

In lid retraction the lower margin of the upper lid is either level with or above the limbus. According to aetiology the condition may be unilateral or bilateral, symmetric or asymmetric, and transient or permanent. The causes are as follows:

Thyroid eye disease

Eponymous signs

a. *Dalrymple* – lid retraction in primary gaze which may be bilateral or unilateral (**Fig. 1.54**).

b. *Kocher* – staring and frightened appearance particularly marked on attentive fixation (**Fig. 1.55**).

c. *von Graefe* – retarded descent of the upper lid on downgaze (lid lag – **Fig. 1.56**).

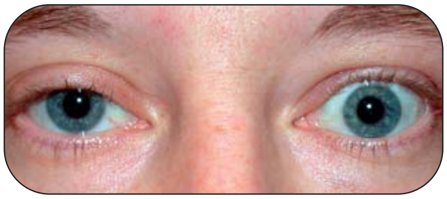

Fig. 1.54

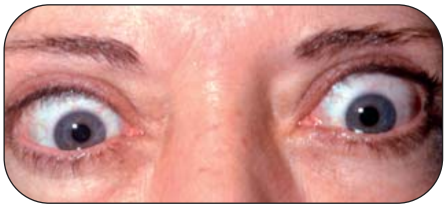

Fig. 1.55

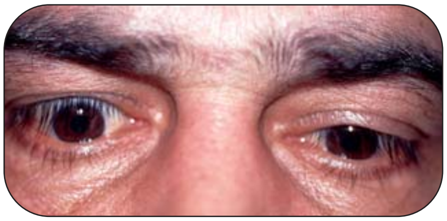

Fig. 1.56

Myasthenia gravis

Myasthenia may cause lid retraction as a result of the following mechanisms:

- Unilateral myasthenic ptosis with contralateral lid retraction caused by compensatory increased innervation to the levator (**Fig. 1.57**).
- Patients with myasthenia may also manifest transient bilateral lid retraction (‘twitch’) as the eyes are brought from downgaze to the primary position.

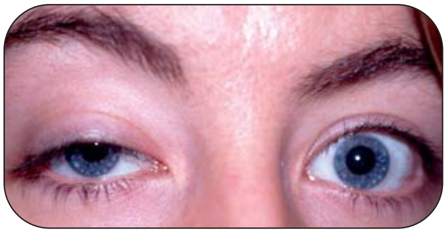

Fig. 1.57

Third nerve misdirection

Definition

- A rare condition that may be congenital or may follow an acquired third nerve palsy.

Signs

- Transient unilateral lid retraction or ptosis induced by various eye movements (**Fig. 1.58**).

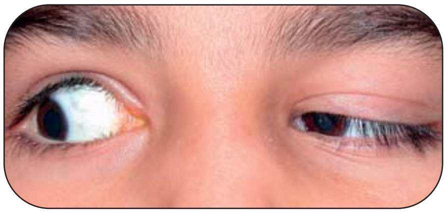

Fig. 1.58

Marcus Gunn jaw-winking syndrome

Definition

- An uncommon congenital, usually unilateral, condition caused by abnormal synkinesis between the levator and the pterygoid muscles.

Signs

- Transient lid retraction may be induced by stimulation of the pterygoid muscles as by opening the mouth (**Fig. 1.59**).
- Less common stimuli to winking include jaw protrusion, smiling, swallowing and clenching of teeth.

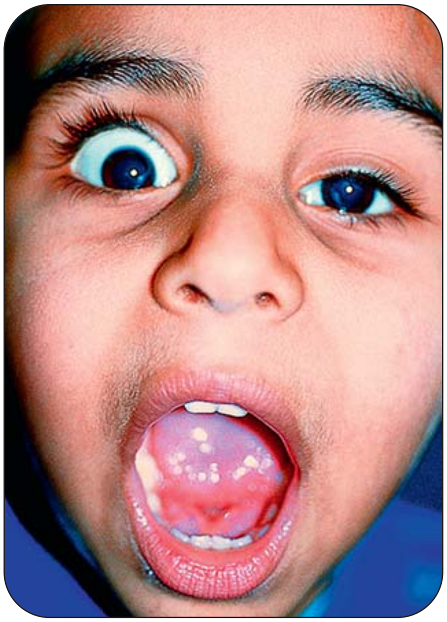

Fig. 1.59

Collier sign of the midbrain (Parinaud syndrome)

Signs

- Bilateral lid retraction is associated with light-near dissociation, impairment of upgaze, convergence deficiency and convergence-retraction nystagmus.

Causes

- See Table 1.6.

Table 1.6 **Causes of Parinaud syndrome**

1. **In children**
   - Pinealoma
   - Aqueduct stenosis
   - Meningitis
2. **In young adults**
   - Demyelination
   - Trauma
   - Arteriovenous aneurysm
3. **In the elderly**
   - Midbrain vascular accidents
   - Mass lesions involving the periaqueductal grey matter
   - Posterior fossa aneurysm

Miscellaneous causes

a. *Infantile hydrocephalus* (setting sun sign – **Fig. 1.60**) in which there is bilateral lid retraction, forceful downward deviation of the eyes and impairment of upgaze.

b. *Facial nerve palsy* with unopposed ipsilateral levator overaction (**Fig. 1.61**).

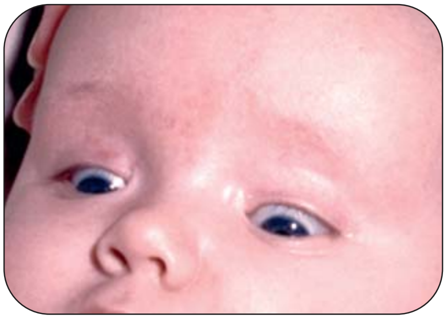

Fig. 1.60

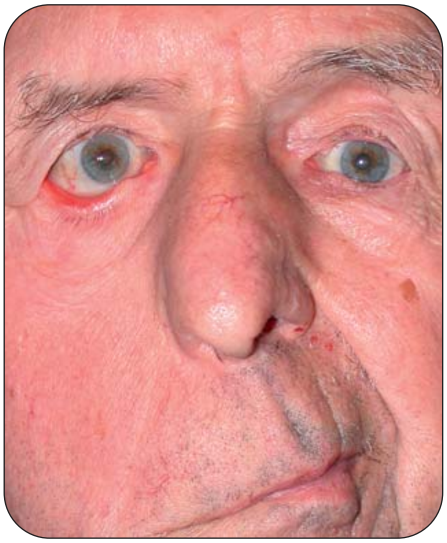

Fig. 1.61

c. *Duane retraction syndrome type 1 and 3* in which there is transient widening of the palpebral fissure and lid retraction on attempted abduction (**Fig. 1.62**).

d. *Parkinsonism* in which there is also lack of facial expression (**Fig. 1.63**), rigidity and tremor.

e. *Isolated congenital* is very rare (**Fig. 1.64**).

f. *Mechanical* may be caused by surgical over-correction of ptosis or scarring of the upper lid.

g. *Down syndrome*.

h. *Transient 'eye popping' reflex* in normal infants.

i. *Uraemia* (Summerskill sign).

j. *Topical sympathomimetic drops*.

k. *Prominent globe* may cause pseudo-lid retraction.

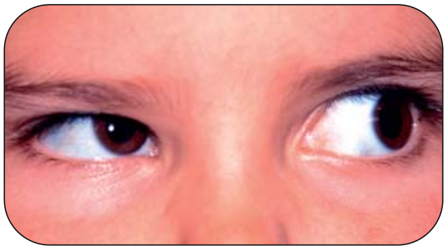

Fig. 1.62

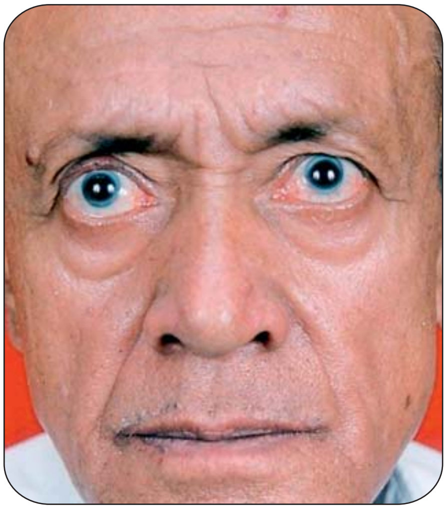

Fig. 1.63

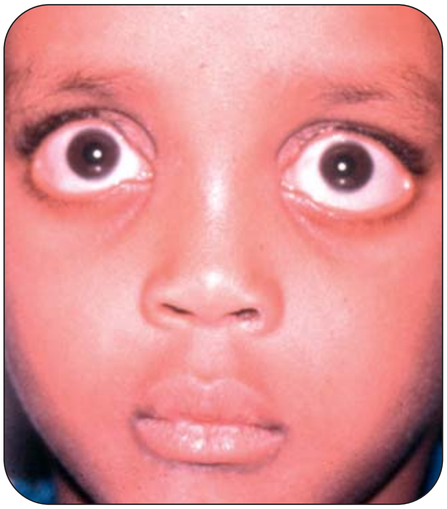

Fig. 1.64

PTOSIS

Neurogenic adult ptosis

Third nerve palsy

Signs

- Ptosis is unilateral and usually moderate to severe (**Fig. 1.65**).
- In the primary position the eye is slightly divergent and depressed.
- Limitation of adduction, elevation and depression.
- Paralysis of accommodation.
- The pupil may or may not be fixed and dilated depending on the cause.

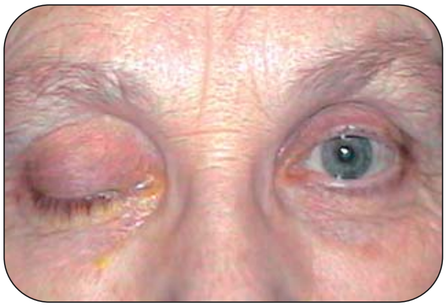

Fig. 1.65

Look for pupillary involvement

- If uninvolved – diabetes and hypertension.
- If involved – posterior communicating aneurysm.

Horner syndrome (oculosympathetic palsy)

Signs

- Mild unilateral ptosis (**Fig. 1.66**).
- Ipsilateral miosis.
- Ipsilateral slight elevation of the lower eyelid.
- Ipsilateral reduction in sweating if the lesion is below the superior cervical ganglion.
- Hypochromic heterochromia iridis may be present if the lesion is congenital or acquired and long-standing.

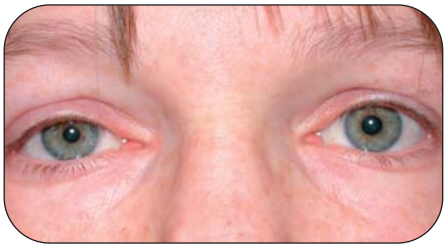

Fig. 1.66

Causes

- See Table 1.7.

Table 1.7 **Causes of Horner syndrome**

1. **Central lesions (first-order neuron)**
    - Brainstem tumour
    - Syringomyelia
    - Lateral medullary (Wallenberg) syndrome
    - Upper spinal cord tumour
2. **Preganglionic lesions (second-order neuron)**
    - Neural crest tumour
    - Neck lesions
    - Intrathoracic lesions (Pancoast tumour, enlarged glands, aneurysm)
3. **Postganglionic lesions (third-order neuron)**
    - Cluster headache (migrainous neuralgia)
    - Nasopharyngeal tumour
    - Otitis media
    - Cavernous sinus mass
    - Internal carotid artery disease
4. **Miscellaneous**
    - Congenital
    - Idiopathic

Third nerve misdirection

Signs

- Transient ptosis or ptosis accompanying various eye movements (see Fig. 1.58).

Myogenic adult ptosis

Myasthenia gravis

Definition

- An autoimmune disease in which antibodies mediate damage and destruction of acetylcholine receptors in striated muscle.
- The three types are ocular, bulbar and generalised.

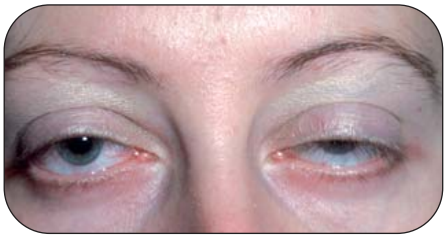

Fig. 1.67

Signs

- Insidious, bilateral but frequently asymmetric ptosis (**Fig. 1.67**), worse with fatigue and in upgaze.
- If one lid is elevated manually as the patient looks up, the fellow lid will show fine oscillatory vertical movements.

- Cogan twitch sign – brief lid retraction as the eyes are brought from downgaze to the primary position.
- Intermittent diplopia, commonly vertical.

Look for systemic

- Excessive muscular fatigue particularly involving the arms and proximal muscles of the legs.
- Problems with speech, chewing and swallowing.
- Thymoma.

Myotonic dystrophy

Definition

- A rare AD condition characterised by delayed relaxation of skeletal muscles after contraction.

Signs

- Bilateral, symmetric ptosis.
- Mournful expression due to facial weakness (**Fig. 1.68**).

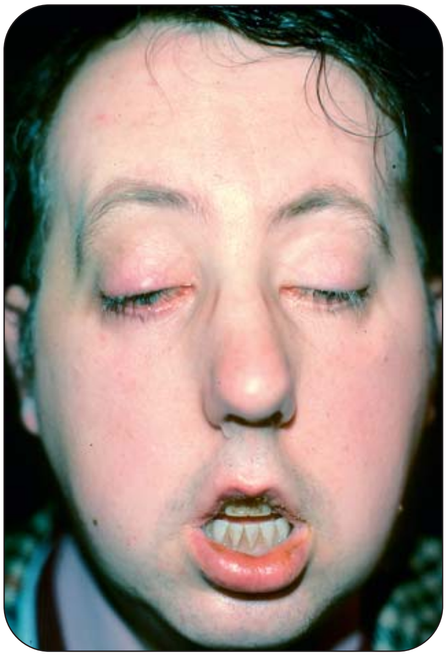

Fig. 1.68

Look for eyes

- Bilateral miosis.
- Light-near dissociation.
- Presenile posterior stellate cataract (see Fig. 10.59).
- Pigmentary retinopathy.
- Mild ophthalmoplegia.
- Low IOP.

Look for systemic

- Myotonia of peripheral muscles resulting in difficulty in relaxing grip.
- Frontal baldness in men.
- Hypogonadism resulting in infertility and sterility.
- Weakness of the sternomastoids.
- Cardiomyopathy and conduction defects.
- Small pituitary fossa.
- Glucose intolerance.

Ocular myopathy

Definition

- A very rare mitochondrial cytopathy.

Signs

- Progressive symmetric ptosis and external ophthalmoplegia (**Fig. 1.69**).

Look for systemic

a. *Oculopharyngeal dystrophy* – weakness of pharyngeal muscles and wasting of the temporalis.

b. *Kearns–Sayre syndrome* – pigmentary retinopathy, heart block and ataxia.

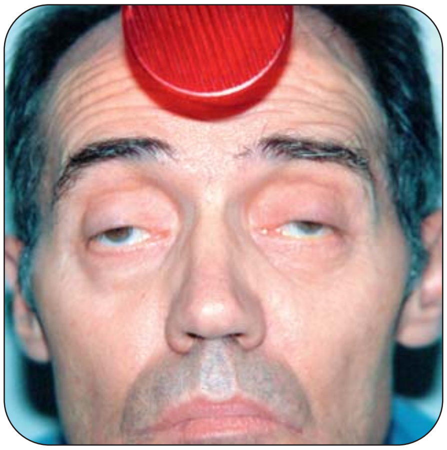

Fig. 1.69

Other types of adult ptosis

Aponeurotic

Signs

- Good levator function.
- High or absent upper-lid crease.
- Thinning of the eyelid above the tarsal plate and deep upper sulci (**Fig. 1.70**).

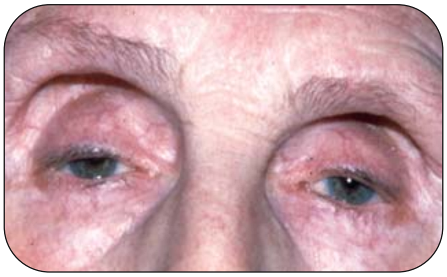

Fig. 1.70

Causes

- Involutional is by far the most common.
- Lid swelling following surgery or blepharochalasis.

Mechanical

Causes

- Severe lid oedema (see Fig. 1.44).
- Eyelid tumours such as plexiform neurofibromas (**Fig. 1.71**).
- Orbital lesions such as cellulitis and anteriorly located tumours (see Fig. 1.46).

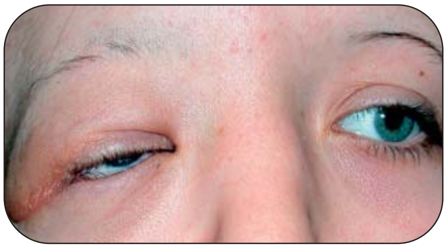

Fig. 1.71

NEUROGENIC CHILDHOOD PTOSIS

Marcus Gunn syndrome

Signs

- Retraction of the ptotic lid in conjunction with stimulation of the pterygoid muscles induced by chewing, sucking, opening the mouth (**Fig. 1.72AB**) or contralateral jaw movements.

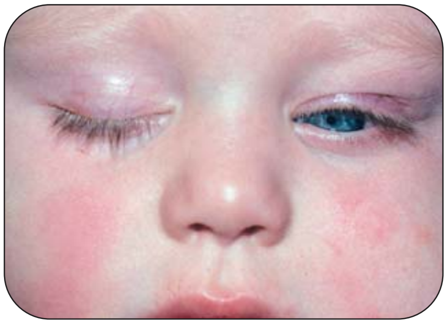

Fig. 1.72A

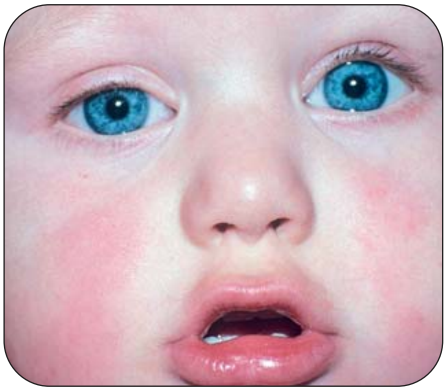

Fig. 1.72B

Horner syndrome (oculosympathetic palsy)

Horner syndrome is rare in children and may be associated with ipsilateral hypochromic heterochromia.

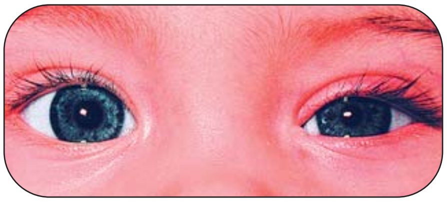

Fig. 1.73

Causes

- Congenital – intrauterine varicella (chickenpox – **Fig. 1.73**), obstetric trauma, although many cases are idiopathic.
- Postnatally acquired – brain stem disease, neck trauma and neuroblastoma.

Third nerve palsy

Causes

- Bilateral congenital – anomalous brain development or prenatal damage.
- Postnatally acquired – trauma, meningitis, tumours and ophthalmoplegic migraine.

Differential diagnosis

- Early-onset esotropia.
- Duane syndrome types 1 and 3.
- Möbius syndrome.

Bassen–Kornzweig syndrome

Definition

- A rare AR disorder of metabolism characterised by fat malabsorption and progressive symmetric ptosis.

Look for eyes

- Ophthalmoplegia.
- Nystagmus.
- Cataracts.
- Progressive retinal dystrophy characterised by white dots and pigmentary retinopathy.

Look for systemic

- Spinocerebellar ataxia.
- Plasma acanthocytosis.
- Hypocholesterolaemia.

Myogenic childhood ptosis

Simple congenital ptosis

Pathogenesis

- Dystrophy of the levator muscle.

Signs

- Unilateral or bilateral ptosis of variable severity (**Fig. 1.74A**).
- Usually poor levator function (**Fig. 1.74B**).
- The ptotic lid is slightly higher in downgaze.
- Ipsilateral weakness of elevation is common.
- Compensatory chin-up position in severe bilateral cases.

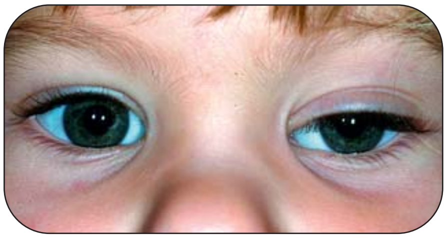

Fig. 1.74A

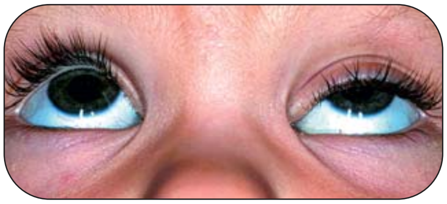

Fig. 1.74B

Blepharophimosis syndrome

Definition

- A rare bilateral congenital AD condition.

Signs

- Moderate–severe symmetric ptosis (**Fig. 1.75**).
- Poor levator function.
- Short horizontal palpebral aperture.
- Epicanthus inversus.
- Lateral ectropion.
- Poorly developed nasal bridge.
- Hypoplasia of the superior orbital rim.
- Telecanthus.

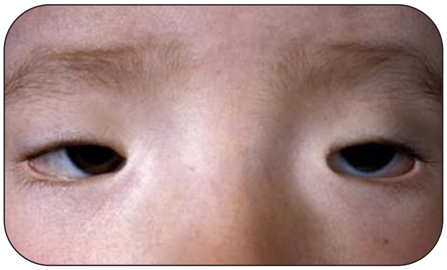

Fig. 1.75

Congenital fibrosis syndrome

Definition

- A very rare non-progressive, usually AD, condition.

Signs

- Severe bilateral congenital ptosis and ophthalmoplegia (**Fig. 1.76**).

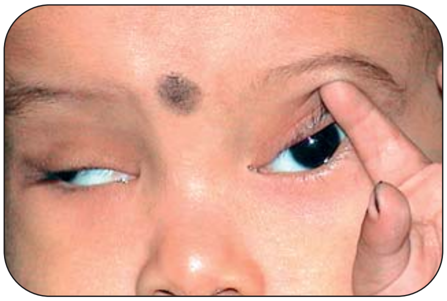

Fig. 1.76

Myasthenia gravis

Myasthenia may rarely affect children (**Fig. 1.77**).

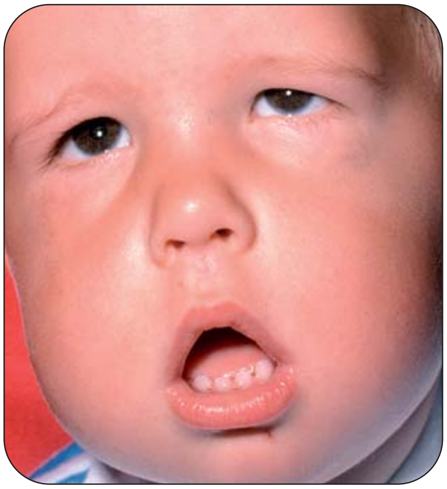

Fig. 1.77

Causes of pseudo-ptosis

In pseudo-ptosis the lid margin is at the normal level but the lid appears ptotic.

Brow ptosis

Causes

- Excessive eyebrow skin or facial nerve palsy.

Signs

- It is diagnosed by manually elevating the eyebrow (**Fig. 1.78**).

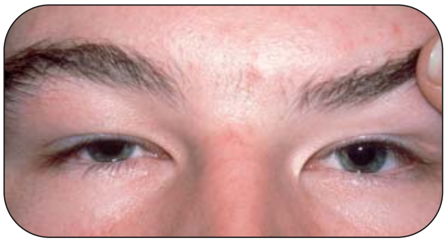

Fig. 1.78

Dermatochalasis

Signs

- Excessive skin on both upper lids (**Fig. 1.79**).

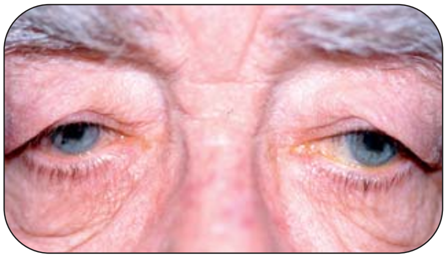

Fig. 1.79

Contralateral lid retraction

Signs

- Detected by comparing the levels of the upper lid margins bearing in mind that the margin of the upper lid normally covers the superior 2 mm of the cornea (**Fig. 1.80**).

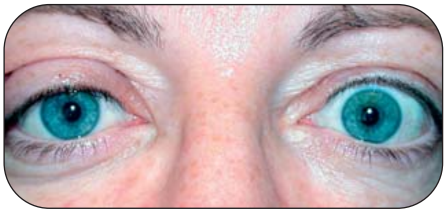

Fig. 1.80

Ipsilateral hypotropia

Signs

- The pseudo-ptosis is ipsilateral to the hypotropia (**Fig. 1.81**).
- It disappears when the hypotropic eye assumes fixation on covering the normal eye.

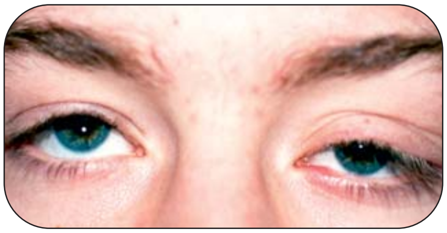

Fig. 1.81

Lack of lid support by the globe

Causes

- Orbital volume deficit that may be associated with microphthalmos, phthisis bulbi, enophthalmos or an ocular prosthesis (**Fig. 1.82**).

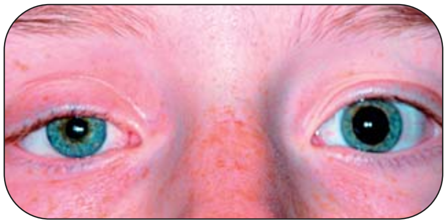

Fig. 1.82

Lash ptosis

Signs

- Downward sagging of upper lashes (**Fig. 1.83**).

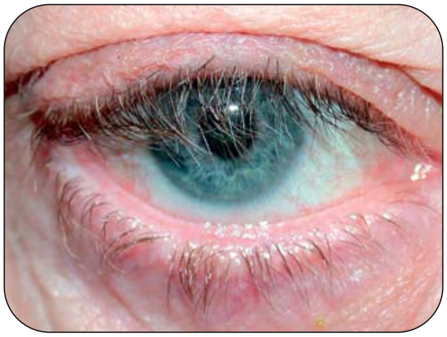

Fig. 1.83

MULTIPLE PAPULES AND VESICLES

Herpes simplex

Signs

- Crops of small vesicles that break down and then resolve after a week (**Fig. 1.84**).

Look for

- Conjunctivitis and keratitis.

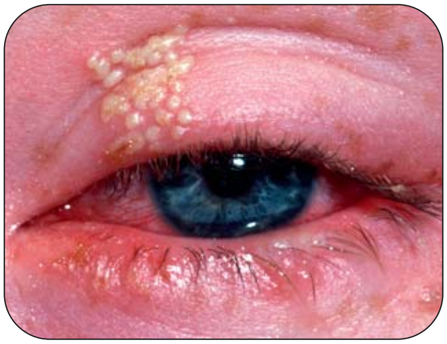

Fig. 1.84

Molluscum contagiosum

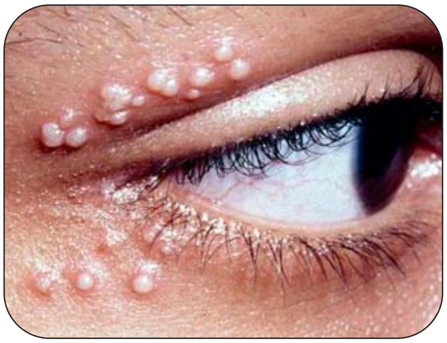
Fig. 1.85

Cause

- A pox virus that typically affects children.

Signs

- Pale waxy umbilicated papules (**Fig. 1.85**).

Look for

- Chronic follicular conjunctivitis with lid margin lesions.
- AIDS in patients with extensive involvement.

Milia

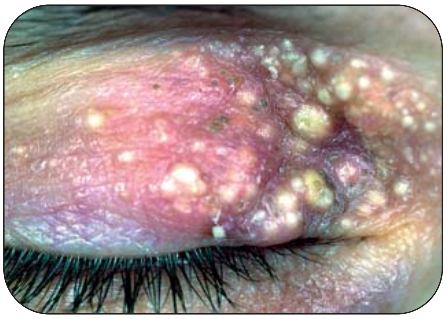
Fig. 1.86

Cause

- Occlusion of pilosebaceous units resulting in retention of keratin.

Signs

- Crops of whitish round papules (**Fig. 1.86**).

Syringomas

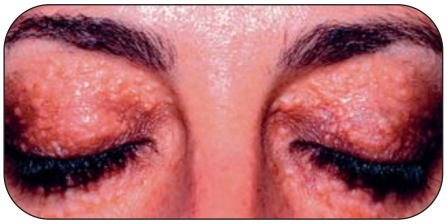
Fig. 1.87

Cause

- Cellular proliferation of intraepidermal duct eccrine sweat gland epithelium.

Signs

- Multiple small papules (**Fig. 1.87**).

Comedones

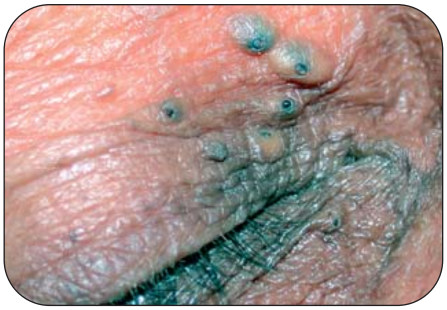
Fig. 1.88

Cause

- Comedones occur in patients with acne vulgaris and consist of a plug of keratin and sebum within the dilated orifice of a hair follicle.

Signs

- Open (blackheads) contain a plug of melanin containing keratin (**Fig. 1.88**).
- Closed (white heads) are cream-coloured papules.

Dermatosis papulosa nigra

Definition

- A variant of seborrhoeic keratosis (see below) that affects black patients.

Signs

- Multiple pigmented papules (**Fig. 1.89**).

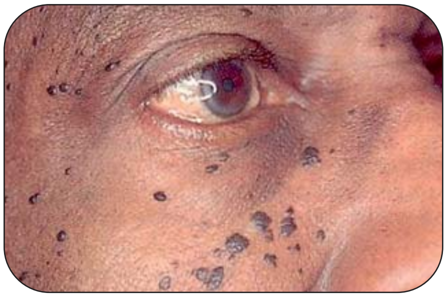

Fig. 1.89

Xanthelasma

Signs

- Bilateral, yellowish, subcutaneous papules at the medial canthi (**Fig. 1.90**).

Look for

- Hypercholesterolaemia.

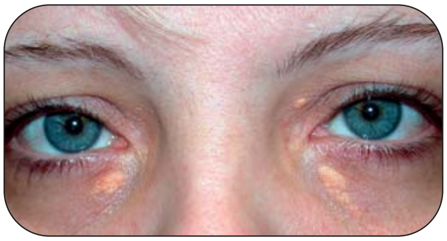

Fig. 1.90

CYSTIC LESIONS

Apocrine sweat gland hidrocystoma (cyst of Moll)

Signs

- Translucent cystic lid margin lesion that contains serous secretions (**Fig. 1.91**).

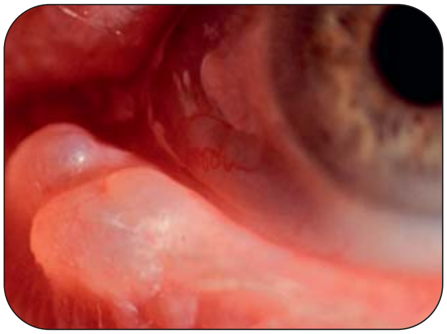

Fig. 1.91

Eccrine sweat gland hidrocystoma

Signs

- Less common but similar to a cyst of Moll except that it is not confined to the lid margin (**Fig. 1.92**).

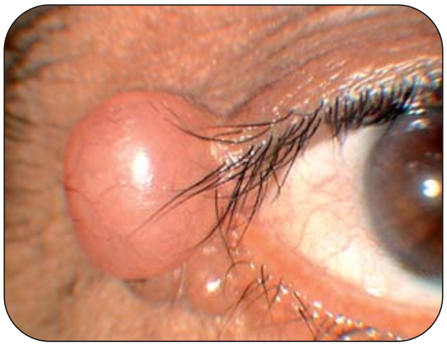

Fig. 1.92

Cyst of Zeis

Signs

- Opaque, smooth cystic lid margin lesion that contains sebaceous secretions (**Fig. 1.93**).

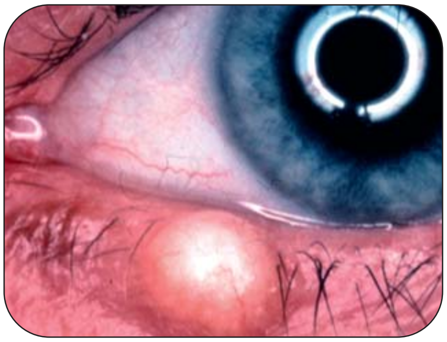

Fig. 1.93

Pilar ('sebaceous') cyst

Signs

- Similar to a cyst of Zeis except that it usually has a central waxy punctum and it most frequently occurs at the inner canthus (**Fig. 1.94**).

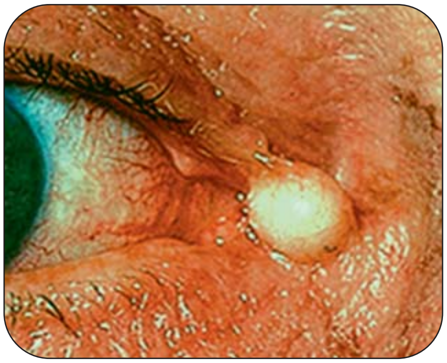

Fig. 1.94

Epidermal inclusion cyst

Cause

- Usually trauma or surgery but may occasionally be congenital.

Signs

- Firm subepithelial nodule which is usually solitary and most commonly located on the upper eyelid (**Fig. 1.95**).

Look for

- Torre syndrome and Gardner syndrome in patients with multiple lesions.

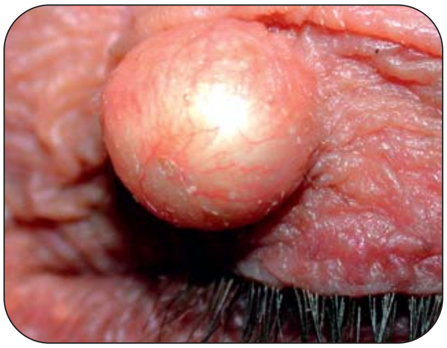

Fig. 1.95

Superficial dermoid cyst

Presentation

- In infancy.

Signs

- Smooth subcutaneous nodule, most frequently below the lateral brow (**Fig. 1.96**).

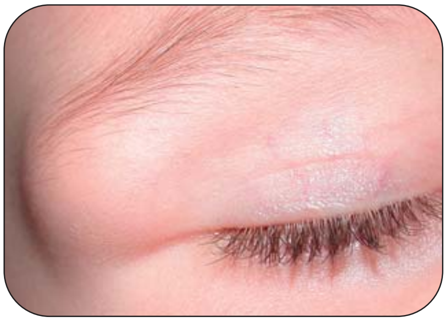

Fig. 1.96

PAPILLOMATOUS LESIONS

Squamous cell papilloma

Signs

- Pedunculated or sessile lesion with a characteristic irregular raspberry-like surface (**Fig. 1.97**).

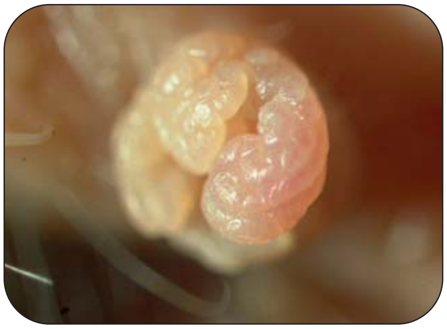

Fig. 1.97

Basal cell papilloma (seborrhoeic keratosis, senile verruca)

Signs

- Discrete, greasy, brown lesion with a friable verrucous surface and a ‘stuck-on’ appearance (**Fig. 1.98**).

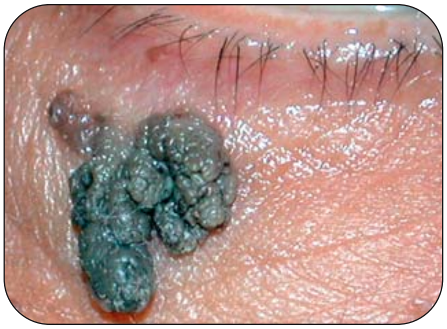

Fig. 1.98

Inverted follicular keratosis

Signs

- Small, fast-growing, wart-like lesion (**Fig. 1.99**).

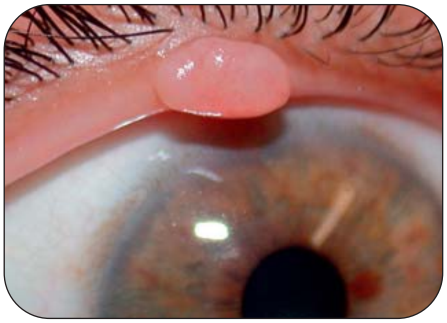

Fig. 1.99

NODULAR LESIONS

Benign

Chalazion (meibomian cyst)

Definition

- A very common chronic lipogranulomatous inflammation of the meibomian glands or glands of Zeis.

Signs

- Painless, roundish, rubbery lesion within the tarsal plate (**Fig. 1.100**).
- May be associated with a conjunctival granuloma (**Fig. 1.101**).

Look for

- Posterior blepharitis.
- Acne rosacea.

Differential diagnosis

- Meibomian gland carcinoma.

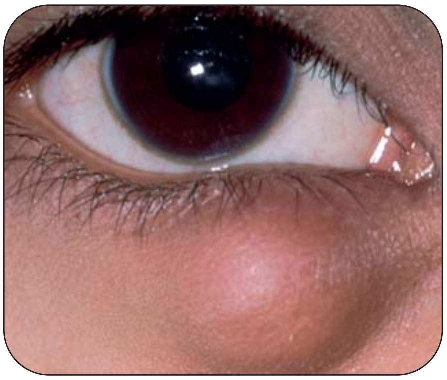

Fig. 1.100

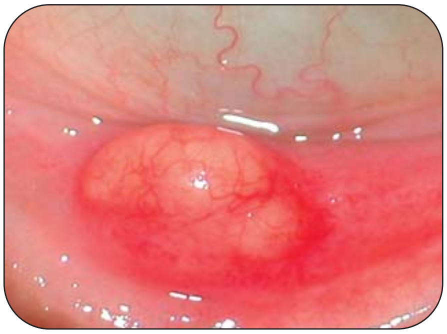

Fig. 1.101

External hordeolum (stye)

Definition

- A common acute purulent infection of a lash follicle and its associated gland of Zeis or Moll.

Signs

- Tender, localised, inflamed swelling in the lid margin, pointing anteriorly through the skin (**Fig. 1.102**).

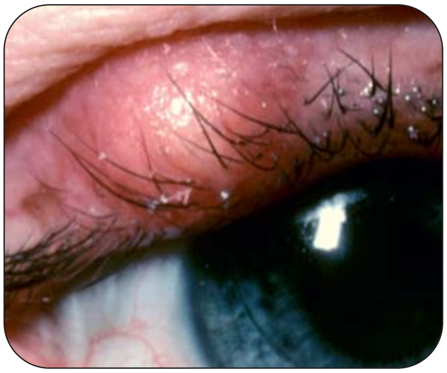

Fig. 1.102

Capillary haemangioma

Presentation

- At birth or early infancy with a predilection for the upper lid.

Signs

- Red nodule that blanches with pressure and may increase in size on crying (**Fig. 1.103**).

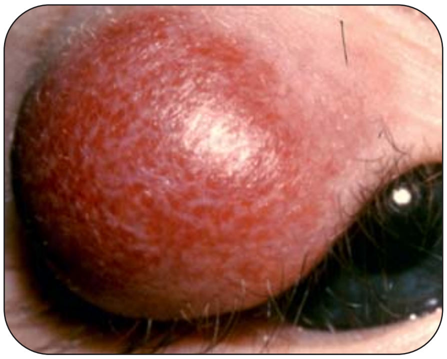

Fig. 1.103

Acquired naevus

Classification

- Acquired naevus can be subdivided into three types according to histology (junctional, compound and dermal).

Signs

- The clinical appearance is determined by the histological location within the skin.
- An intradermal naevus (most common) is a non-pigmented nodule through which lashes may protrude (**Fig. 1.104**).

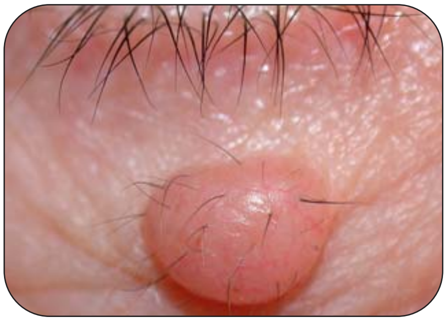

Fig. 1.104

Actinic (solar, senile) keratosis

Definition

- A very slow-growing condition that typically affects elderly, fair-skinned individuals exposed to excessive sunlight. It predisposes to squamous cell carcinoma.

Signs

- Hyperkeratotic nodule (**Fig. 1.105**) or plaque.

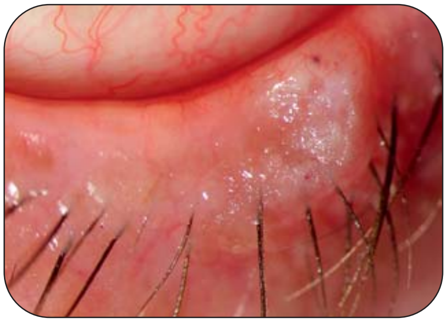

Fig. 1.105

Keratoacanthoma

Definition

- A fast-growing tumour that typically affects fair-skinned individuals with a history of chronic sun exposure.

Signs

- Starts as a firm, pinkish, indurated nodule which quickly acquires a dome-shaped configuration and a keratin-filled crater (**Fig. 1.106**).

Differential diagnosis

- Squamous cell carcinoma.

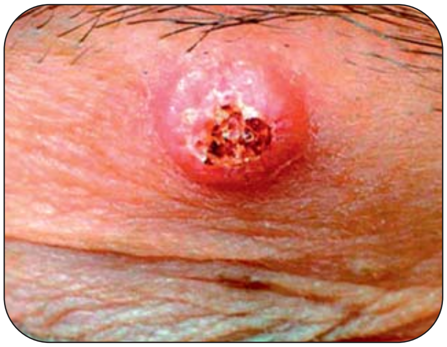

Fig. 1.106

Neurofibroma

Definition

- A rare benign tumour that typically affects patients with NF1.

Signs

- Large subcutaneous nodule that causes a characteristic S-shaped deformity of the upper lid (**Fig. 1.107**).

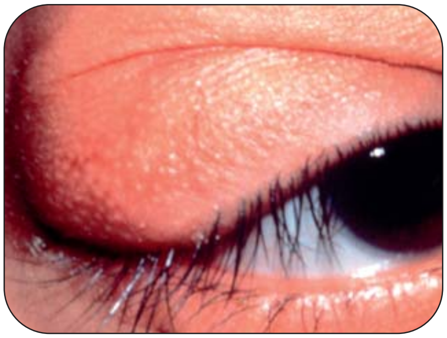

Fig. 1.107

Pilomatricoma (pilomatrixoma)

Definition

- The most common hair follicular proliferation that typically affects young individuals.

Signs

- Purplish, mobile, subcutaneous nodule that is often hard due to calcification (**Fig. 1.108**).

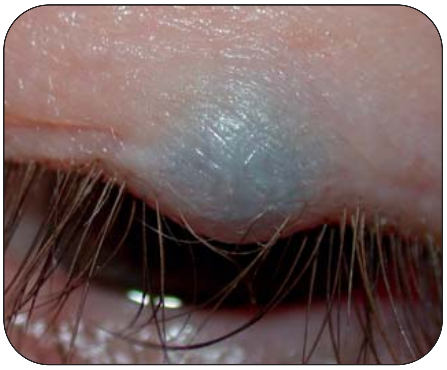

Fig. 1.108

Trichilemmoma

Definition

- An uncommon tumour that arises from the outer sheath of a hair follicle and typically affects older individuals.

Signs

- Small nodular (**Fig. 1.109**) or papillomatous lesion.

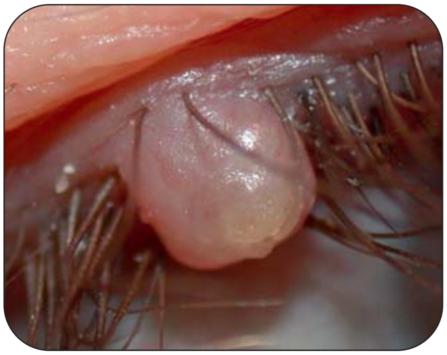

Fig. 1.109

Cutaneous horn

Signs

- Hyperkeratotic, horn-like lesion protruding from the skin surface (**Fig. 1.110**).

Look for

- Associated actinic keratosis or squamous cell carcinoma.

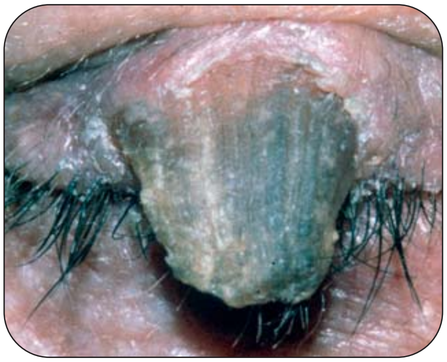

Fig. 1.110

Malignant

Nodular basal cell carcinoma

Signs

- Slow-growing, firm, shiny, indurated nodule with fine surface telangiectasia (**Fig. 1.111**).

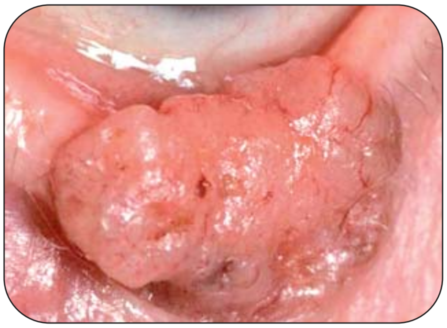

Fig. 1.111

Nodular squamous cell carcinoma

Signs

- Hyperkeratotic nodule with crusting erosions and fissures (**Fig. 1.112**) but unlike basal cell carcinoma there is absence of telangiectasia.
- Grows faster than a basal cell carcinoma.

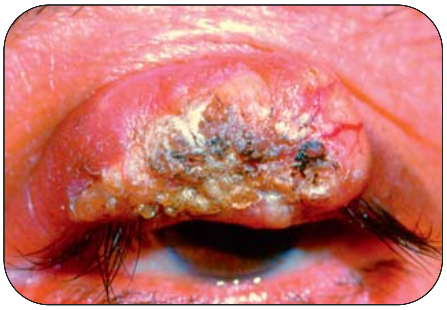

Fig. 1.112

Nodular meibomian gland carcinoma

Signs

- Hard nodule most commonly within the upper tarsal plate often associated with loss of lashes (**Fig. 1.113**).

Differential diagnosis

- Chalazion is more rubbery and localised.

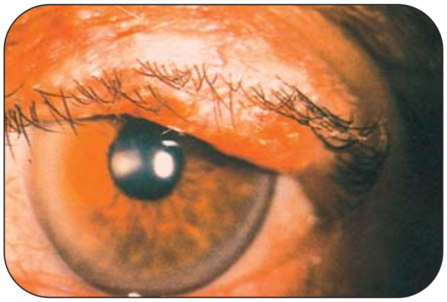

Fig. 1.113

Kaposi sarcoma

Signs

- Red-violet nodule that may be solitary or multiple (**Fig. 1.114**).

Look for

- AIDS.

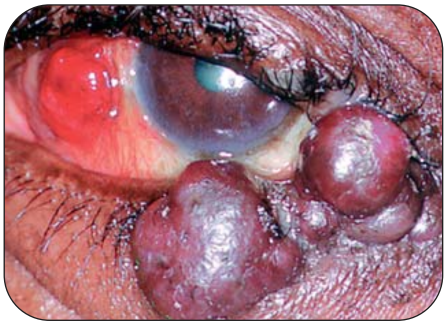

Fig. 1.114

Nodular melanoma

Signs

- Irregularly-shaped nodule with variable pigmentation that may grow rapidly (**Fig. 1.115**).

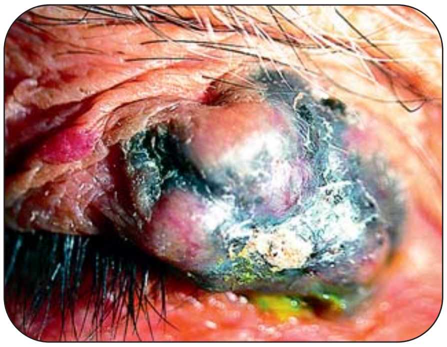

Fig. 1.115

Merkel cell (neuroendocrine) carcinoma

Definition

- A very rare aggressive tumour that occurs in elderly individuals.

Signs

- Fast-growing, red or purple nodule with overlying telangiectasia (**Fig. 1.116**).

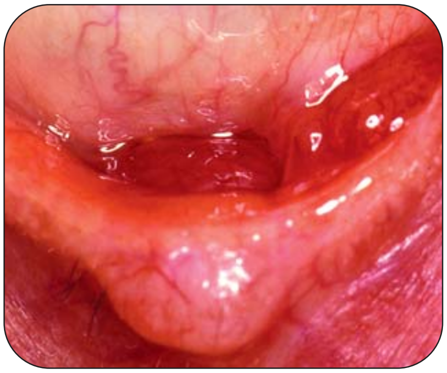

Fig. 1.116

ULCERATING TUMOURS

Ulcerating basal cell carcinoma (rodent ulcer)

Signs

- Rolled edges with pearly margins and surface telangiectasia (**Fig. 1.117**).

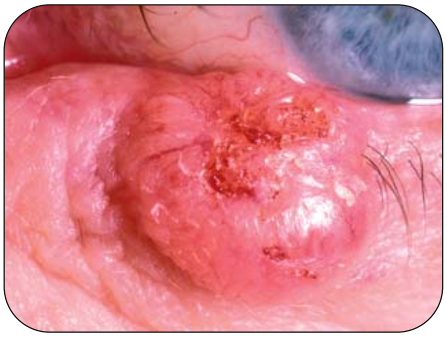

Fig. 1.117

Ulcerating squamous cell carcinoma

Signs

- Red base with sharply defined, indurated and everted borders with surface crusting but without telangiectasia (**Fig. 1.118**).

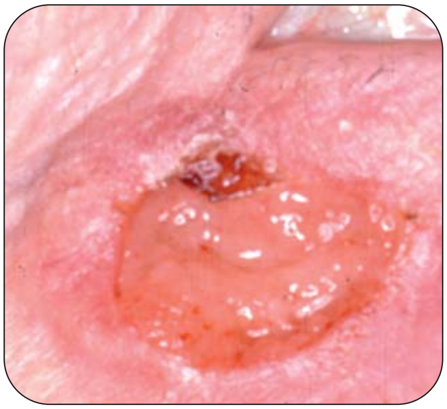

Fig. 1.118

Ulcerating gland of Zeis carcinoma

Signs

- Yellow ulcerating nodule on the lid margin (**Fig. 1.119**).

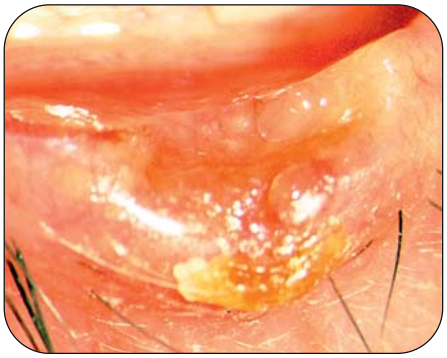

Fig. 1.119

Table 1.8 **Ulcerating BCC vs SCC**

	BCC	**SCC**
Incidence	common	rare
Growth rate	slow	faster
Metastatic potential	minimal	considerable
Pearly margins	++++	nil
Surface telangiectasia	++++	nil
Rolled edges	yes	no
Surface crusting	nil	+++
Cutaneous horn formation	nil	+++
Associated with actinic keratosis and Bowen disease	no	yes
Most commonly on lower lid	+++	++

PLAQUE-LIKE LESIONS

Bowen disease (intraepidermal carcinoma *in situ*)

Signs

- Red scaling plaque that may be mistaken for a patch of psoriasis (**Fig. 1.120**).

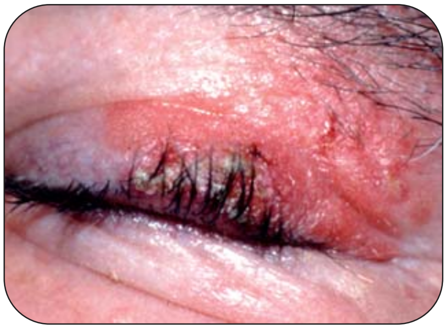

Fig. 1.120

Sclerosing basal cell carcinoma

Signs

- Indurated plaque with poorly demarcated margins and an intact epidermis often associated with destruction of the overlying lashes (**Fig. 1.121**).

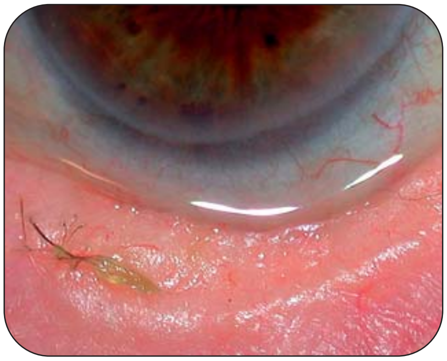

Fig. 1.121

Spreading meibomian gland carcinoma

Signs

- Similar in appearance to sclerosing basal cell tumour but is usually on the upper lid (**Fig. 1.122**).
- The tumour may also spread within the epithelium of the conjunctiva (pagetoid spread) and mimic non-specific chronic conjunctivitis (see Fig. 3.52).

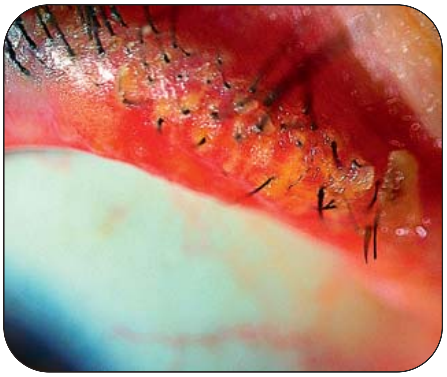
Fig. 1.122

Superficial spreading melanoma

This may arise from lentigo maligna (Hutchinson freckle).

Signs

- Plaque with irregular outline and variable pigmentation (**Fig. 1.123**).
- May arise from lentigo maligna (see Fig. 1.49).

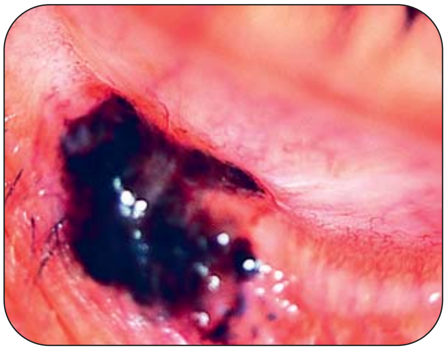
Fig. 1.123

MAJOR CONGENITAL ANOMALIES

Ablepharon

Cause

- Congenital deficiency of the anterior lamellae of the eyelids.

Look for

- Enlarged fish-like mouth, ear and genital anomalies, and redundant skin (ablepharon-macrostomia syndrome – **Fig. 1.124**).

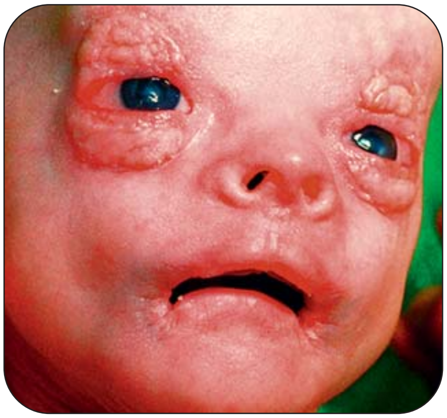
Fig. 1.124

Microblepharon

Signs

- Small or rudimentary lids often associated with anophthalmos (**Fig. 1.125**).

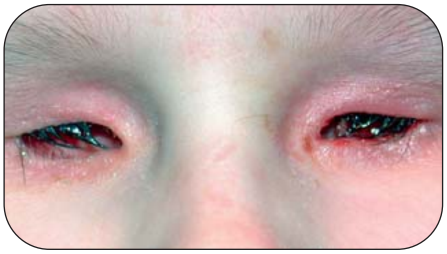
Fig. 1.125

Coloboma

Definition

- An uncommon, bilateral or unilateral, partial or full-thickness defect involving the lid margin.

Signs

- Upper lid coloboma occurs at the junction of the inner and middle thirds (**Fig. 1.126**) and is often associated with Goldenhar syndrome.
- Lower lid coloboma occurs at the junction of the middle and outer thirds and may be associated with Treacher Collins syndrome and midfacial clefts (**Fig. 1.127**).

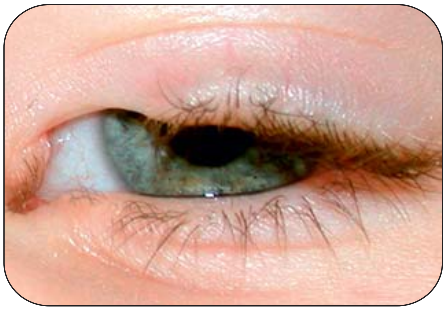

Fig. 1.126

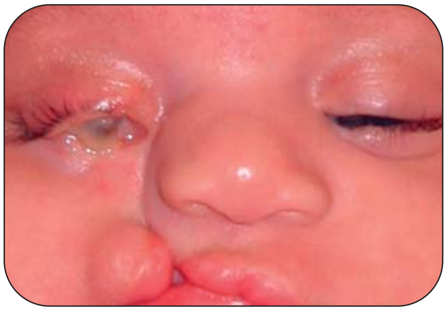

Fig. 1.127

Congenital lid inversion

Definition

- A very rare, bilateral condition that may be associated with Down syndrome or collodion skin disease which is a type of ichthyosis (**Fig. 1.128**).

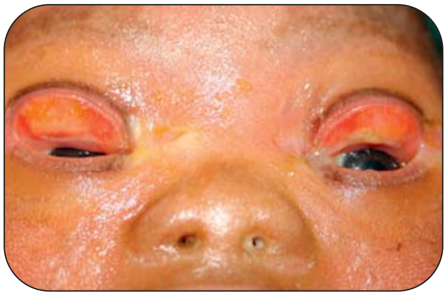

Fig. 1.128

Cryptophthalmos

Signs

- The lids are replaced by a fold of skin which is fused with a microphthalmic eye.
- The condition may be complete (**Fig. 1.129**) or partial (**Fig. 1.130**); the latter is associated with Fraser syndrome.

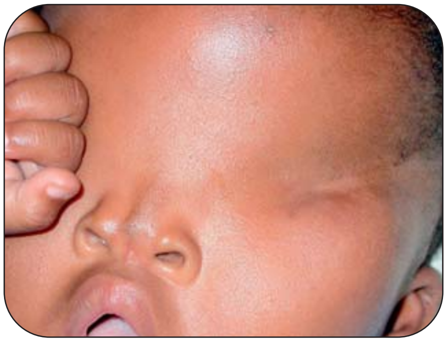

Fig. 1.129

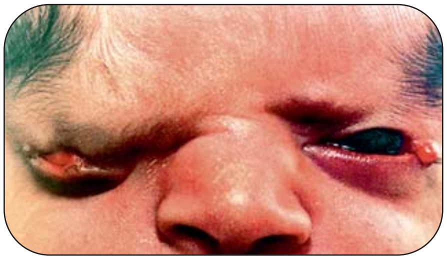

Fig. 1.130

Orbit 2

CLINICAL SIGNS IN ORBITAL DISEASE 41

BONY ABNORMALITIES 41

Hypertelorism 41

Dystopia 42

Shallow orbits associated with craniosynostoses 42

Oxycephaly 42

Crouzon syndrome 43

Apert syndrome 43

Pfeiffer syndrome 44

Other causes 44

ENOPHTHALMOS 44

Small globe 44

Structural bony abnormalities 45

Atrophy of orbital contents 45

Cicatrising orbital lesions 45

Duane retraction syndrome 45

PROPTOSIS 45

ORBITAL DISEASE IN CHILDREN 46

Inflammatory 46

Preseptal cellulitis 46

Orbital cellulitis 46

Idiopathic orbital inflammatory syndrome (pseudo-tumour) 46

Thyroid eye disease 47

Benign tumours 47

Capillary haemangioma 47

Lymphangioma 47

Plexiform neurofibroma 48

Optic nerve glioma 48

Langerhans-cell histiocytosis 49

Other benign tumours 49

Malignant tumours 49

Embryonal sarcoma (rhabdomyosarcoma) 49

Metastatic neuroblastoma 50

Myeloblastoma (granulocytic granuloma) 50

Other malignant tumours 50

Cystic lesions 51

Deep dermoid cyst 51

Microphthalmos with cyst 51

Anophthalmos with cyst (congenital cystic eyeball) 51

Anterior orbital encephalocele 51

Posterior orbital encephalocele 52

ORBITAL DISEASE IN ADULTS 52

Congestive proptosis 52

Thyroid eye disease 52

Carotid–cavernous fistula 53

Cavernous sinus thrombosis 53

Metastatic tumour 53

Other causes 53

Chronic non-congestive axial proptosis 54

Thyroid eye disease 54

Cavernous haemangioma 54

Optic nerve sheath meningioma 54

Orbital varices 55

Chronic non-congestive non-axial proptosis 55

Lymphoid tumours 55

Sphenoidal ridge meningioma 56

Mucocele 56

Orbital extension of sinus tumours 56

Other causes 57

LACRIMAL GLAND ENLARGEMENT 57

Unilateral 57

Dacryoadenitis 57

Pleomorphic adenoma (benign mixed cell tumour) 58

Lacrimal gland carcinoma 58

Dacryops 59

Bilateral 59

ORBITAL RIM LESIONS 59

DYNAMIC PROPTOSIS 60

Intermittent 60

Pulsatile 60

CLINICAL SIGNS IN ORBITAL DISEASE

1. Bony

- Hypertelorism.
- Dystopia in which the orbits are not level.
- Shallow orbits.

2. Soft tissue

- Lid oedema.
- Ptosis.
- Chemosis.

3. Globe malposition

- Proptosis.
- Enophthalmos.
- Displacement.

4. Ophthalmoplegia

- By an orbital mass.
- Restrictive myopathy.
- Ocular motor nerve palsy.
- Muscle entrapment in a fracture.

5. Posterior segment

- Disc oedema.
- Optic atrophy.
- Chorioretinal folds.
- Retinal vascular congestion.

BONY ABNORMALITIES

Hypertelorism

Signs

- Wide separation of the orbits (**Fig. 2.1**).

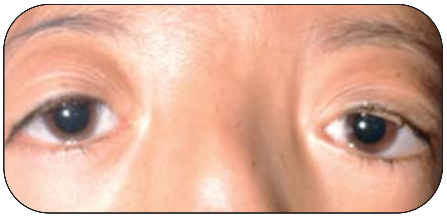

Fig. 2.1

Causes

- Facial trauma.
- Associated with congenital systemic abnormalities (see below).

Differential diagnosis

- Telecanthus in which there is increased distance between the medial canthi as a result of abnormally long medial canthal tendons.

Look for systemic

- Craniosynostosis (see below).
- Frontonasal dysplasia.
- Basal encephalocele with wide nasal bridge.
- Syndromic conditions – Hurler, Noonan, Roberts, Gorlin–Goltz and Meckel–Gruber.

Dystopia

Signs

- Orbits are not level.

Causes

- See Table 2.1.

Table 2.1 **Causes of orbital dystopia**

1. **Congenital**
   - Idiopathic
   - Goldenhar syndrome
   - Hemifacial microsomia
   - Coronal craniosynostosis
   - Facial clefting syndromes (**Fig. 2.2**)
2. **Acquired**
   - Fractures of orbital floor or rim
   - Fibrous dysplasia
   - Meningioma en plaque

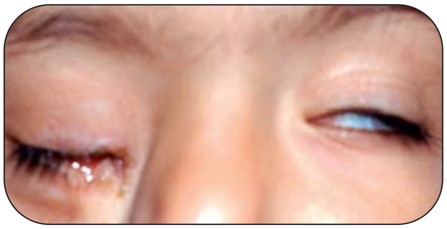

Fig. 2.2

Shallow orbits associated with craniosynostoses

Oxycephaly

Signs

- High, narrow, pointed skull, high forehead and superior prognathism (**Fig. 2.3**).

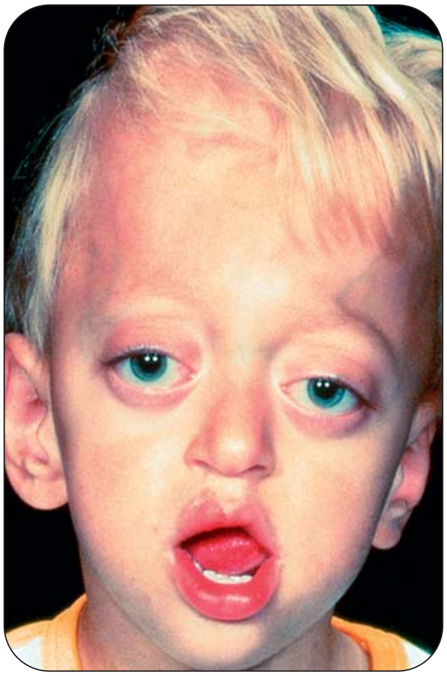

Fig. 2.3

Crouzon syndrome

Signs

- Midfacial hypoplasia, orbital axes are rotated laterally (exorbitism), flat nasal bridge, parrot's beak nose, prominent lower jaw and V pattern exotropia (**Fig. 2.4**).

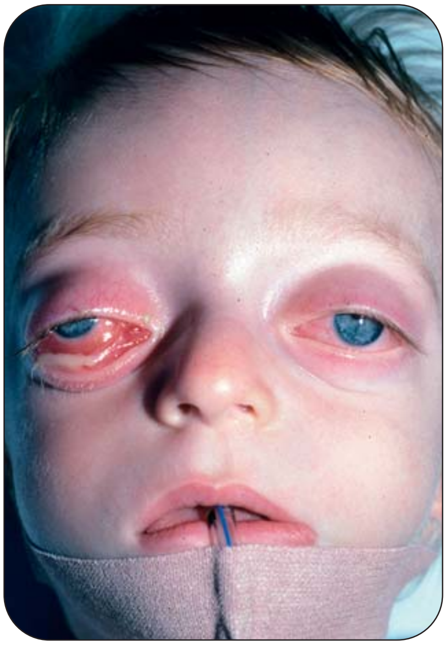

Fig. 2.4

Apert syndrome

Signs

- Similar to Crouzon but with less severe proptosis and hypertelorism (**Fig. 2.5**).

Look for

- Broad thumbs and great toes, and syndactyly.

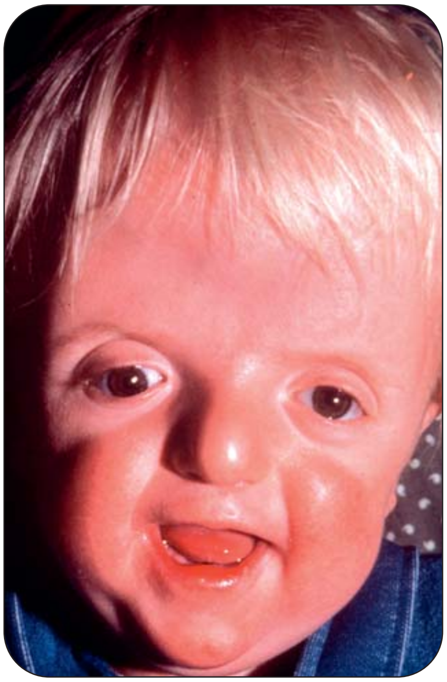

Fig. 2.5

Pfeiffer syndrome

Signs

- Acrocephaly (peaked skull), midfacial hypoplasia, orbital rotation and down-sloping palpebral fissures (**Fig. 2.6**).

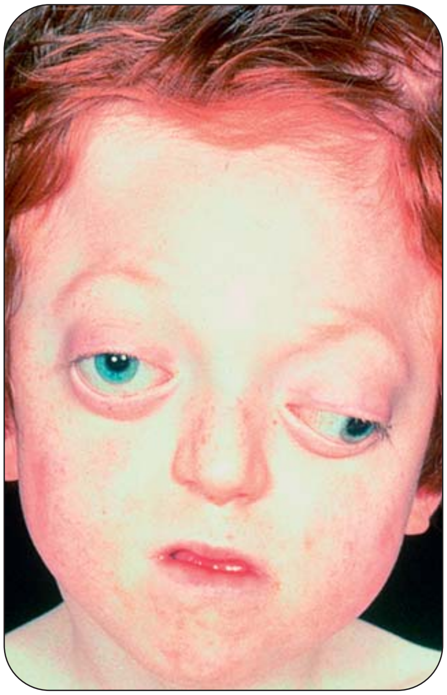

Fig. 2.6

Other causes

a. *Chromosomal abnormalities*

- Trisomy 13 (Patau syndrome).
- Trisomy 18 (Edward syndrome).

b. *Other bony defects*

- Osteogenesis imperfecta.
- Osteopetrosis.
- Hyperphosphataemia.

ENOPHTHALMOS

In enophthalmos the globe is recessed within the orbit. It may be caused by the following conditions.

Small globe

a. *Phthisis bulbi* (**Fig. 2.7**).

b. *Microphthalmos* in which the net volume of the globe is reduced; it may be simple (**Fig. 2.8**) or colobomatous.

c. *Nanophthalmos* which is characterised by a small eye, high hypermetropia, weak but thick sclera, predisposition to angle closure glaucoma and uveal effusion.

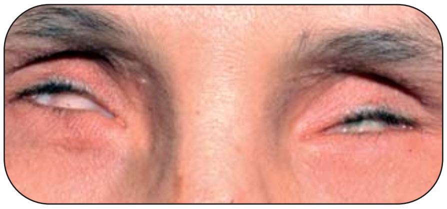

Fig. 2.7

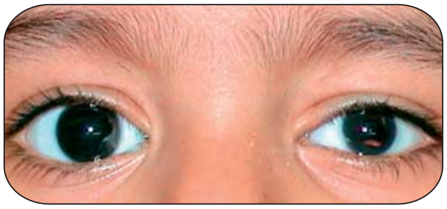

Fig. 2.8

Structural bony abnormalities

a. *After blow-out fracture of orbital floor* (**Fig. 2.9**).

b. *Congenital bony defects* as in NF1.

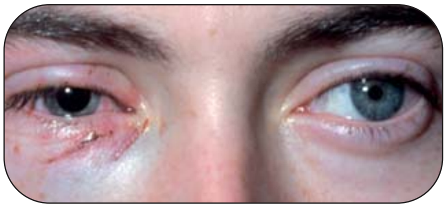

Fig. 2.9

Atrophy of orbital contents

a. *Following radiotherapy.*

b. *Parry–Romberg hemifacial atrophy* (see Fig. 8.95).

c. *Scleroderma.*

d. *Chronic maxillary sinusitis.*

e. *Long-standing orbital varices* (**Fig. 2.10**).

f. *Eye poking (oculo-digital sign)* in blind children (**Fig. 2.11**).

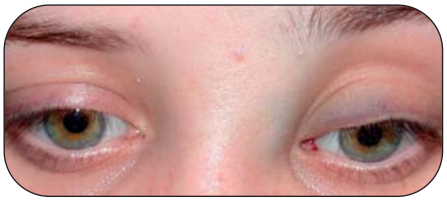

Fig. 2.10

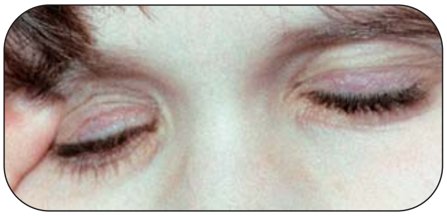

Fig. 2.11

Cicatrising orbital lesions

a. *Metastatic scirrhous breast carcinoma.*

b. *Chronic sclerosing inflammatory orbital disease*.

Duane retraction syndrome

Although this is usually characterised by retraction of the globe on adduction rarely there may be true enophthalmos in the primary position (see Fig. 14.17A).

PROPTOSIS

Signs

- The globe is pushed forward – best detected by looking down from above (**Fig. 2.12**).

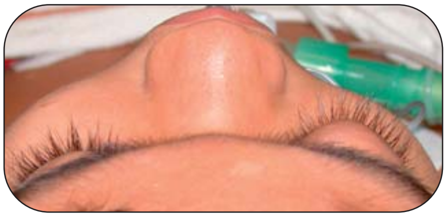

Fig. 2.12

Direction

- Axial proptosis is caused by lesions within the muscle cone such as cavernous haemangioma, optic nerve tumours and thyroid eye disease.
- Eccentric proptosis is caused by extraconal lesions in which the direction of proptosis is determined by the site of the lesion.

Causes of pseudo-proptosis

- Ipsilateral large globe as in buphthalmos (**Fig. 2.13**) or very high myopia.
- Ipsilateral lid retraction.
- Contralateral enophthalmos.

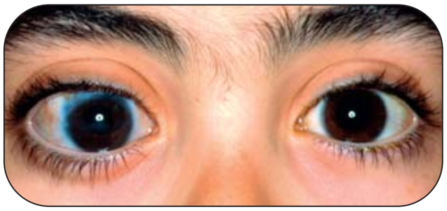

Fig. 2.13

ORBITAL DISEASE IN CHILDREN

Inflammatory

Preseptal cellulitis

Definition

- An infection of the subcutaneous tissues anterior to the orbital septum.

Signs

- Unilateral, tender and red periorbital oedema (**Fig. 2.14A**).
- Normal ocular motility and visual acuity.
- CT shows opacification anterior to the orbital septum (**Fig. 2.14B**).

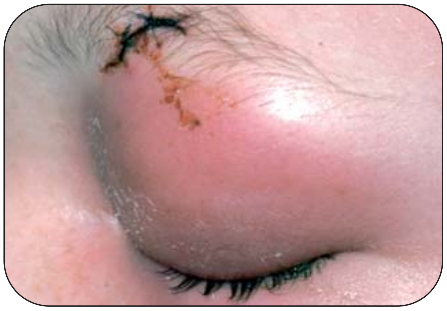

Fig. 2.14A

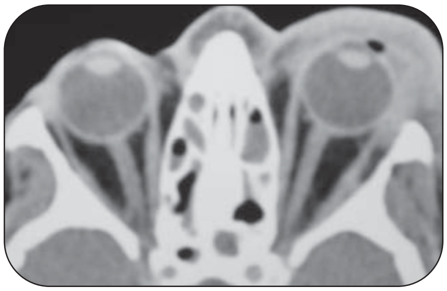

Fig. 2.14B

Orbital cellulitis

Definition

- A life-threatening infection of soft tissues posterior to the orbital septum, which in children, is usually secondary to ethmoiditis.

Signs

- Very unwell patient with a high temperature.
- Rapid onset of painful proptosis (usually down and out), chemosis, lid oedema and ophthalmoplegia (**Fig. 2.15A**).
- Optic nerve dysfunction in severe cases.
- CT shows preseptal and orbital opacification (**Fig. 2.15B**).

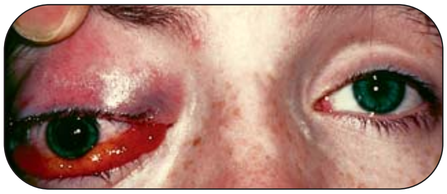

Fig. 2.15A

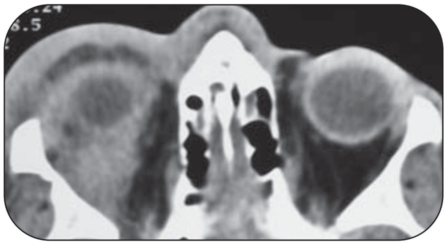

Fig. 2.15B

Idiopathic orbital inflammatory syndrome (pseudo-tumour)

Definition

- An uncommon, non-neoplastic and non-infectious space-occupying condition that may present in childhood or adult life. Acute orbital myositis is a subtype which primarily affects one or more of the extraocular muscles.

Signs

- Subacute onset of proptosis associated with lid oedema and displacement of the globe (**Fig. 2.16A**).
- In acute orbital myositis, pain and diplopia are increased on attempted gaze into the field of the affected muscle.
- CT shows a poorly-defined opacification with ill-defined orbital structures (**Fig. 2.16B**).

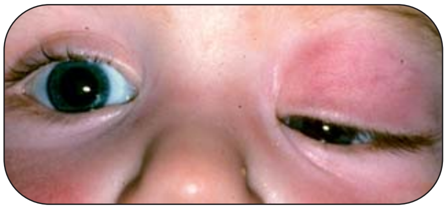

Fig. 2.16A

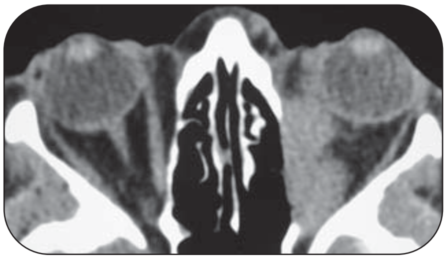

Fig. 2.16B

Look for systemic disease in bilateral cases

- Wegener granulomatosis.
- Polyarteritis nodosa.
- Sarcoidosis.
- Tuberculosis.
- Waldenström macroglobulinaemia.

Thyroid eye disease

Thyroid eye disease may rarely present in children as young as 10 years and cause proptosis and lid retraction (**Fig. 2.17**).

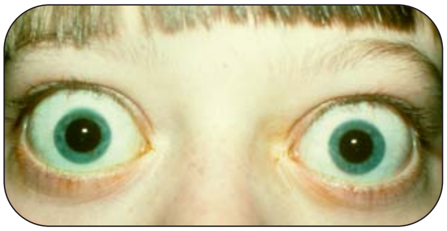

Fig. 2.17

Benign tumours

Capillary haemangioma

Presentation

- At birth or early infancy.

Signs

- Anterior orbit lesion that may appear dark blue or purple through overlying skin and may be associated with cutaneous lesions (**Fig. 2.18**).
- The lesion may become engorged and slightly increases in size when the child cries.

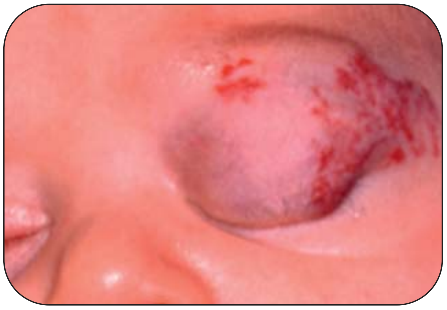

Fig. 2.18

Lymphangioma

Presentation

- Between the ages of 1 and 15 years.

Signs

- Most commonly located in the superior orbit.
- Visual compromise and extraocular motility impairment (**Fig. 2.19A**) are more common than with capillary haemangioma.

Look for

- Involvement of the sinuses and oropharynx (**Fig. 2.19B**).

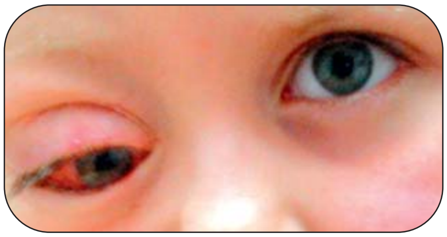

Fig. 2.19A

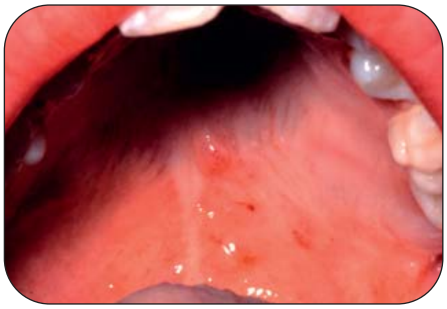

Fig. 2.19B

Plexiform neurofibroma

Presentation

- Between the ages of 2 and 5 years.

Signs

- Non-axial proptosis which may be associated with eyelid or facial hypertrophy (**Fig. 2.20**).
- The proptosis is pulsatile if there is an associated congenital defect in the sphenoid bone.

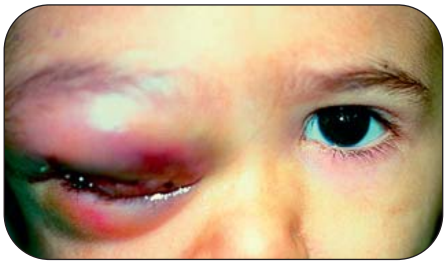

Fig. 2.20

Look for

- NF1.

Optic nerve glioma

Presentation

- Between the ages of 2 and 7 years.

Signs

- Slowly progressive axial proptosis (**Fig. 2.21A**) associated with decreased visual acuity and an afferent pupillary conduction defect.
- The optic disc may show swelling, atrophy, or opticociliary shunts (see Fig. 12.45).
- There is a disproportionate loss of visual acuity compared to the severity of proptosis.
- CT shows a fusiform enlargement of the optic nerve with a clear-cut margin corresponding to the dural sheath (**Fig. 2.21B**).

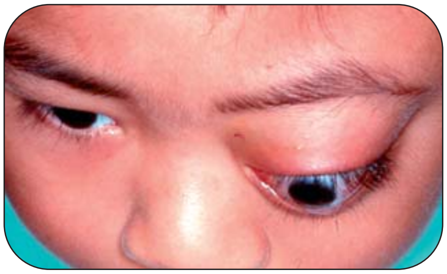

Fig. 2.21A

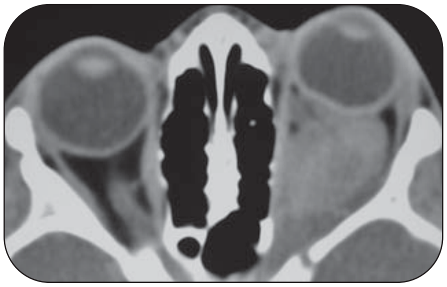

Fig. 2.21B

Look for

- NF1, which is present in 50% of cases with unilateral tumours and in 100% with bilateral involvement.

Langerhans-cell histiocytosis

Definition

- A rare multisystem disorder characterised by proliferation of abnormal histiocytes.

Signs

- Bilateral or unilateral bony lysis and soft-tissue proliferation which typically involves the superotemporal orbit (**Fig. 2.22**).

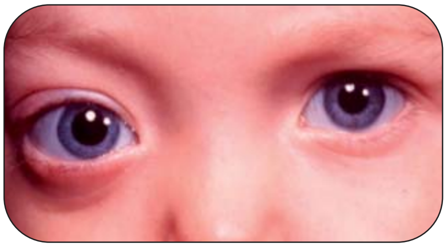

Fig. 2.22

Other benign tumours

a. *Sinus histiocytosis* is characterised by massive painless cervical lymphadenopathy and extranodal involvement of other viscera, with bilateral or unilateral orbital involvement.

b. *Teratoma* presents at birth with massive proptosis.

c. *Deep dermoid cyst* presents in adolescence with non-axial proptosis. It may occasionally rupture and cause inflammation.

d. *Fibrous dysplasia* typically involves the orbital roof and causes proptosis with downward displacement of the globe. It may be associated with McCune–Albright syndrome.

e. *Juvenile ossifying fibroma* arises from either the roof or the ethmoids. It causes chronic painless proptosis with downward or lateral globe displacement.

f. *Osteopetrosis* is a hereditary condition characterised by increased density and thickness of bone with increased susceptibility to fracture. It causes optic atrophy and bilateral proptosis due to shallow orbits.

Malignant tumours

Embryonal sarcoma (rhabdomyosarcoma)

Definition

- A rare but aggressive tumour derived from undifferentiated mesenchymal cell rests that typically affects boys at around the age of 7 years.

Signs

- Rapid onset of progressive painful proptosis associated with chemosis and lid oedema and erythema (**Fig. 2.23A**).
- The most common location is retrobulbar, followed by superior and inferior.
- CT shows a moderately well-defined homogeneous mass that is isodense with extraocular muscles (**Fig. 2.23B**).

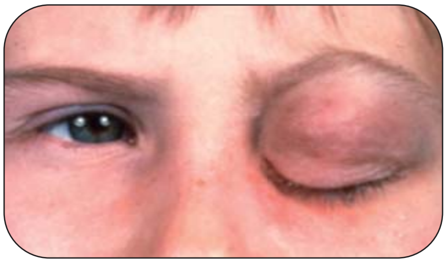

Fig. 2.23A

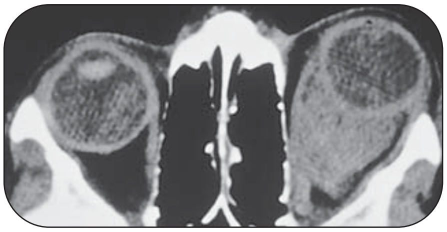

Fig. 2.23B

Metastatic neuroblastoma

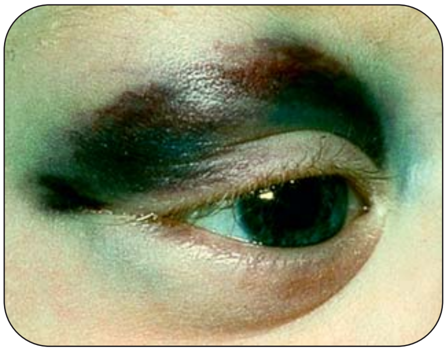
Fig. 2.24

Definition

- An aggressive tumour that presents during the first 5 years of life.
- The primary tumour usually develops in the abdomen.

Signs

- Rapid onset proptosis associated with lid ecchymosis which is bilateral in 40% of cases (**Fig. 2.24**).

Myeloblastoma (granulocytic granuloma)

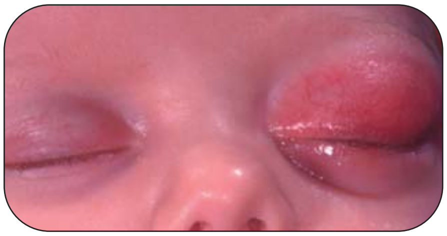
Fig. 2.25

Definition

- A localised form of acute myeloblastic leukaemia that has a predilection for the orbit. It may occasionally precede systemic manifestations.

Signs

- Rapid onset of proptosis, sometimes bilateral.
- Eyelid ecchymosis and oedema (**Fig. 2.25**).

Other malignant tumours

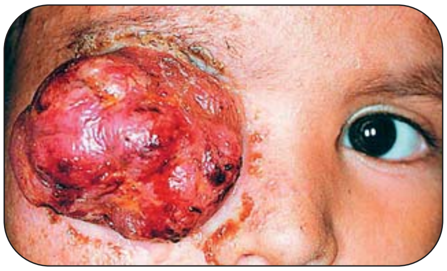
Fig. 2.26

a. *Neglected retinoblastoma* may occasionally present with orbital involvement (**Fig. 2.26**), although occasionally the tumour may incite an orbital inflammatory response mimicking cellulitis.

b. *Metastatic Ewing sarcoma* may cause rapid proptosis, ecchymosis and chemosis.

c. *Metastatic Wilms tumour* which may be associated with aniridia.

d. *Fibrosarcoma* typically occurs in children who have previously received radiotherapy to the orbit for retinoblastoma.

e. *Sinus tumour* invading the orbit such as Burkitt lymphoma and osteogenic sarcoma.

Cystic lesions

Deep dermoid cyst

Signs

- Gradual-onset proptosis and/or globe displacement (**Fig. 2.27A**).
- Occasionally the cyst may rupture and incite an acute inflammatory reaction.
- CT shows a well-circumscribed cystic lesion (**Fig. 2.27B**).

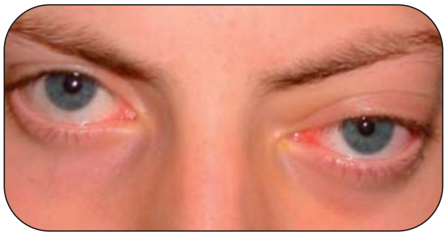

Fig. 2.27A

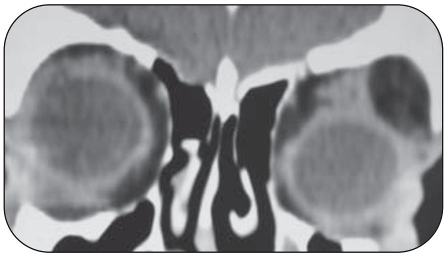

Fig. 2.27B

Microphthalmos with cyst

Pathogenesis

- Incomplete closure of the fetal fissure leading to the formation of an orbital cyst that communicates with the eye. It is best demonstrated on CT (**Fig. 2.28**).

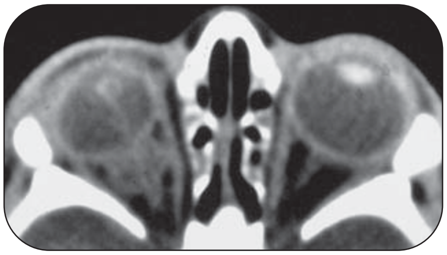

Fig. 2.28

Anophthalmos with cyst (congenital cystic eyeball)

Signs

- One or both globes are replaced by a cyst (**Fig. 2.29A** and **B**).

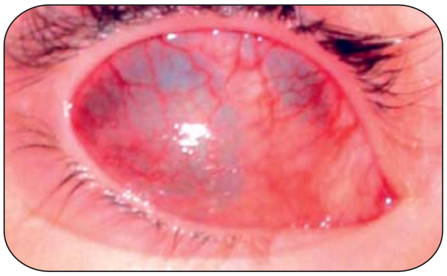

Fig. 2.29A

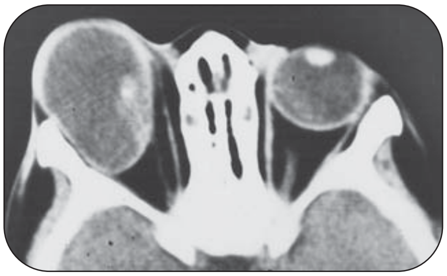

Fig. 2.29B

Anterior orbital encephalocele

Pathogenesis

- A congenital fronto-ethmoidal orbital defect.

Signs

- Proptosis is slowly progressive, pulsatile, increases in size on crying or straining, and is associated with lateral displacement of the globe (**Fig. 2.30**).

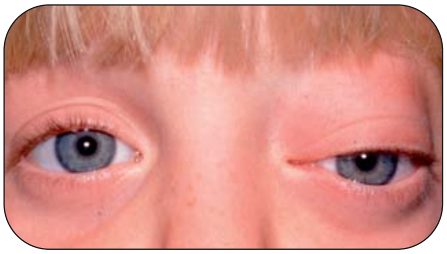

Fig. 2.30

Posterior orbital encephalocele

Pathogenesis

- Dysplasia of the sphenoid bone that may be associated with NF1.

Signs

- Similar to the anterior variety except that the globe is displaced downwards (**Fig. 2.31**).

Table 2.2 **Causes of rapid proptosis in children**

1. **Non-malignant**
    - Orbital cellulitis
    - Ruptured deep dermoid cyst
    - Chocolate cyst with lymphangioma
    - Idiopathic orbital inflammatory disease
2. **Malignant**
    - Rhabdomyosarcoma
    - Acute leukaemia
    - Metastases

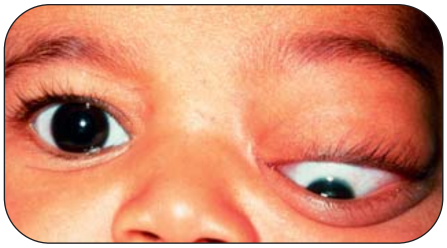

Fig. 2.31

ORBITAL DISEASE IN ADULTS

Congestive proptosis

Congestive proptosis is characterised by a rapid onset of lid oedema, chemosis, conjunctival congestion, ocular motility restriction and, frequently, pain.

Thyroid eye disease

Signs

- Proptosis is axial, unilateral (**Fig. 2.32A**) or bilateral, and is associated with lid retraction or lid lag.
- CT shows proptosis and enlargement of extraocular muscles with absence of space between the muscles and the optic nerve in the posterior orbit (**Fig. 2.32B**).

Look for

- Injection over horizontal recti (see Fig. 3.56).
- Superior limbic keratoconjunctivitis (see Fig. 5.47).
- Restrictive ocular motility defects (see Fig. 14.40).
- Signs of optic nerve compression.
- Chorioretinal folds (see Fig. 13.284).

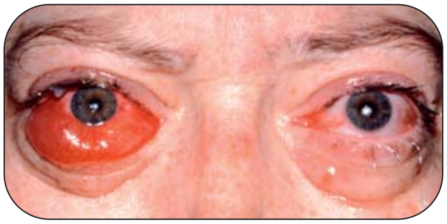

Fig. 2.32A

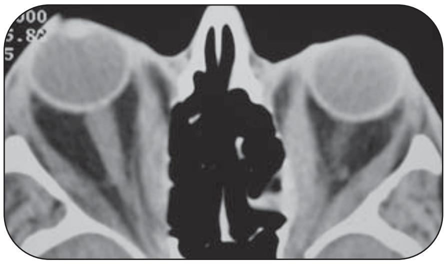

Fig. 2.32B

Carotid–cavernous fistula

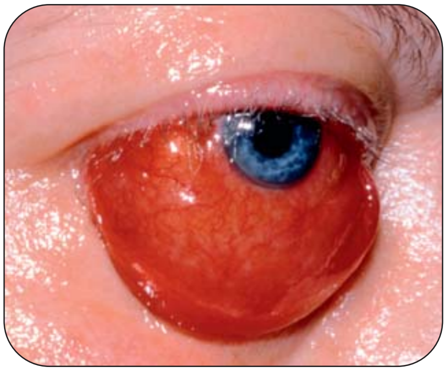

Fig. 2.33

Cause

- Head trauma or a spontaneous rupture of an intracavernous aneurysm.

Signs

- Unilateral, painful, pulsatile proptosis associated with a bruit and severe chemosis (**Fig. 2.33**).

Look for

- Grossly dilated epibulbar vessels may be present in the absence of chemosis (see Fig. 3.54).
- Ophthalmoplegia.
- Retinopathy.

Cavernous sinus thrombosis

Cause

- Skin or paranasal sinus infection.

Signs

- Similar to a carotid–cavernous fistula except that the patient is usually more ill because of systemic infection.

Metastatic tumour

Signs

- Rapid onset of diplopia, lid oedema and pain.
- Scirrhous breast carcinoma may give rise to enophthalmos.

Other causes

a. *Orbital cellulitis* (see above).

b. *Idiopathic orbital inflammatory syndrome* (see above).

Chronic non-congestive axial proptosis

Thyroid eye disease

Signs

- Unilateral or bilateral proptosis associated with eyelid signs (**Fig. 2.34**).

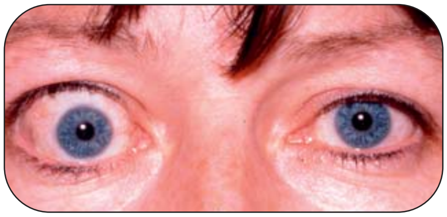

Fig. 2.34

Cavernous haemangioma

Signs

- Unilateral, slowly progressive proptosis (**Fig. 2.35A**).
- Optic nerve compression is uncommon unless the tumour is located near the orbital apex.
- Acquired hypermetropia and chorioretinal folds.
- CT shows an oval well-circumscribed intraconal lesion (**Fig. 2.35B**).

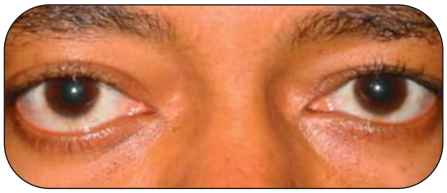

Fig. 2.35A

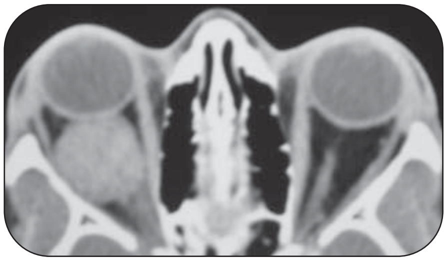

Fig. 2.35B

Optic nerve sheath meningioma

Presentation

- Typically in middle-aged women.

Signs

- Early decrease in visual acuity.
- Opticociliary shunts are common (**Fig. 2.36**).
- CT shows thickening and calcification of the optic nerve (**Fig. 2.37**).

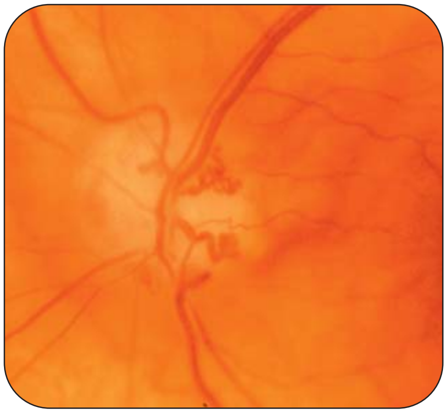

Fig. 2.36

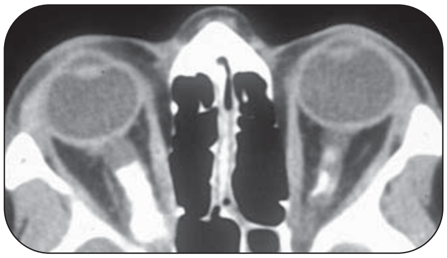

Fig. 2.37

Orbital varices

Presentation

- At any age with intermittent, non-pulsatile proptosis not associated with a bruit.

Signs

- Proptosis may be induced or accentuated by performing the Valsalva manoeuvre (**Fig. 2.38A** before Valsalva, **Fig. 2.38B** after Valsalva).
- CT shows a poorly-defined opacification which may be associated with phleboliths (**Fig. 2.39**).

Look for

- Varices in the eyelids (**Fig. 2.40A**) and conjunctiva (**Fig. 2.40B**).

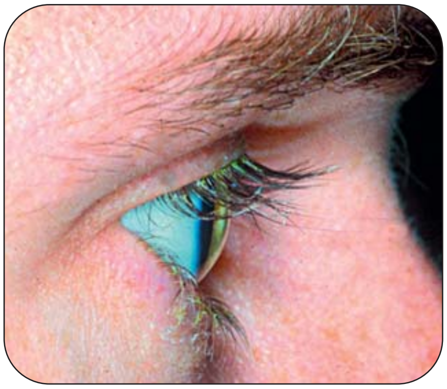
Fig. 2.38A

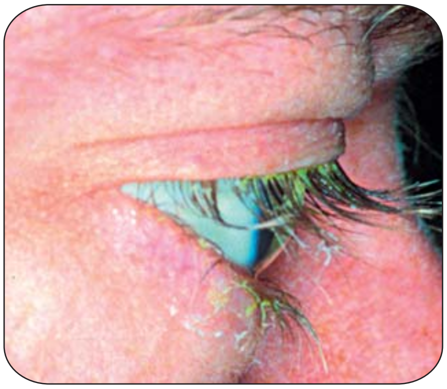
Fig. 2.38B

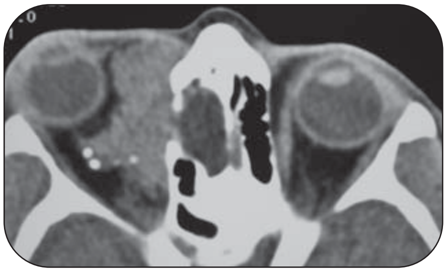
Fig. 2.39

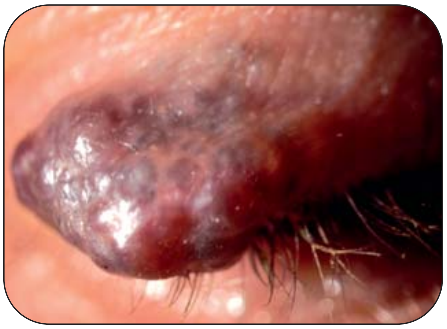
Fig. 2.40A

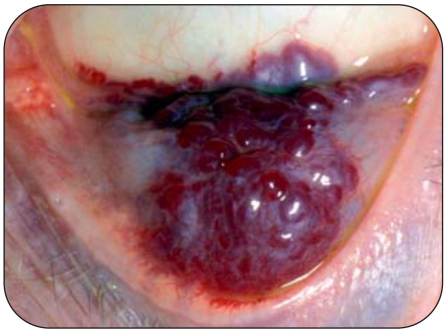
Fig. 2.40B

Chronic non-congestive non-axial proptosis

Non-axial proptosis is caused by anterior orbital disorders which displace the globe away from the lesion.

Lymphoid tumours

Presentation

- Usually in old age.
- May involve any part of one or both orbits.

Signs

- Anterior tumours may displace the globe and cause periocular puffiness (**Fig. 2.41**).

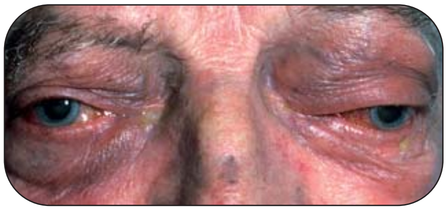
Fig. 2.41

Sphenoidal ridge meningioma

Presentation

- In middle age with slowly progressive, painless proptosis.

Signs

- Lateral globe displacement and periocular swelling (**Fig. 2.42A**).
- Fullness of the temporal fossa may be seen.
- CT shows hyperostosis (**Fig. 2.42B**).

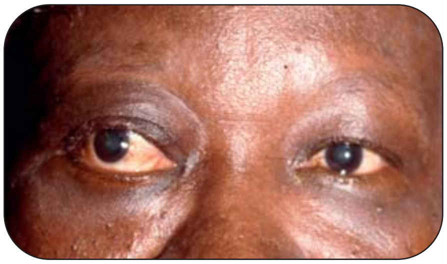
Fig. 2.42A

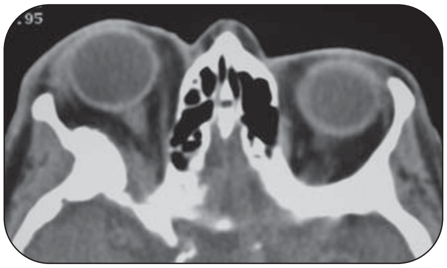

Fig. 2.42B

Mucocele

Presentation

- With a combination of ptosis, proptosis and globe displacement.
- The proptosis may fluctuate when the walls of the mucocele become inflamed.

Signs

a. *Frontal* mucocele displaces the globe downwards and may cause impairment of upgaze (**Fig. 2.43A**). CT shows a bony dehiscence in the orbital roof (**Fig. 2.43B**).

b. *Ethmoidal* mucocele causes lateral displacement which may be associated with proptosis (**Fig. 2.44A**). CT shows loss of ethmoidal septae and erosion into the orbit (**Fig. 2.44B**).

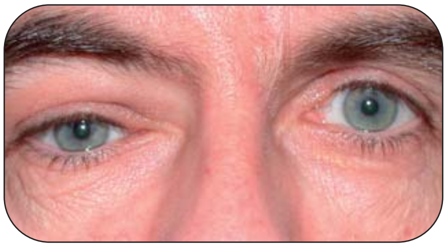
Fig. 2.43A

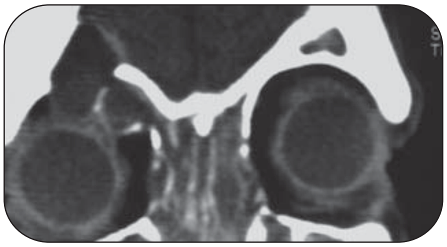
Fig. 2.43B

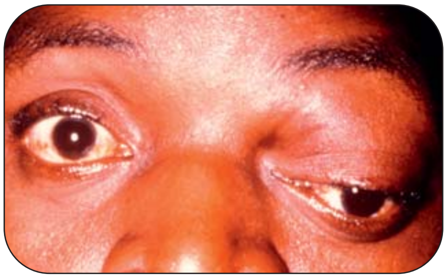
Fig. 2.44A

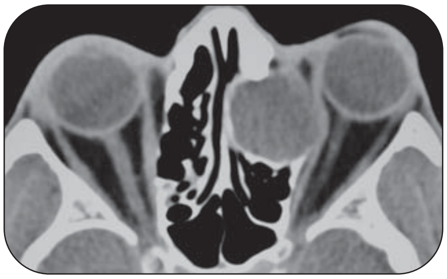
Fig. 2.44B

Orbital extension of sinus tumours

a. *Ethmoidal/frontal carcinoma* displaces the globe down and out.

b. *Maxillary carcinoma* displaces the globe up (**Fig. 2.45**) and is frequently associated with pain and epiphora.

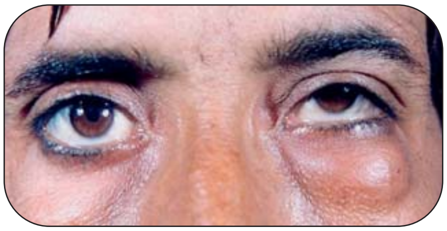
Fig. 2.45

Other causes

a. *Malignant eyelid tumours* may invade the orbit if neglected.

b. *Nasopharyngeal carcinoma* may invade the orbit through the ethmoidal sinus and displaces the globe down and out.

c. *Myeloma* may involve the orbit as a solitary bony deposit (plasmocytoma) or as part of generalised disease.

d. *Leukaemia* may involve the orbit in adults usually as part of established systemic disease.

e. *Lacrimal gland lesions* (see below).

LACRIMAL GLAND ENLARGEMENT

Unilateral

Dacryoadenitis

Definition

- An uncommon condition that may be infectious or may be associated with idiopathic orbital inflammatory disease.

Signs

- The upper lid shows a typical 'S'-shaped curve with erythema of the skin (**Fig. 2.46A**).
- The gland itself feels gritty and there is injection of the gland and adjacent conjunctiva (**Fig. 2.46B**).
- CT shows opacification that extends beyond the gland itself (**Fig. 2.46C**).

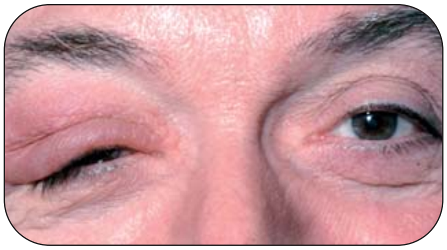

Fig. 2.46A

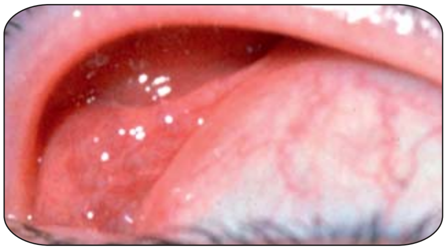

Fig. 2.46B

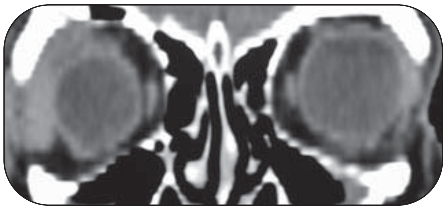

Fig. 2.46C

Pleomorphic adenoma (benign mixed cell tumour)

Presentation

- In middle age with a long painless history.

Signs

- Fullness of the upper eyelid and displacement of the globe down and in (**Fig. 2.47A**).
- A tumour of the palpebral lobe of the lacrimal gland may be visible to inspection (**Fig. 2.47B**) and does not cause displacement of the globe.
- CT shows an oval mass with a smooth outline that may indent the globe and lacrimal fossa (**Fig. 2.47C**).

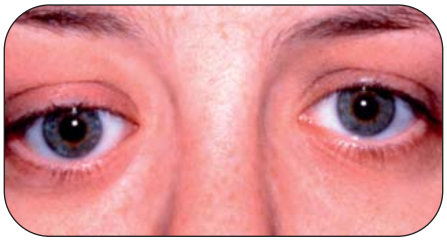
Fig. 2.47A

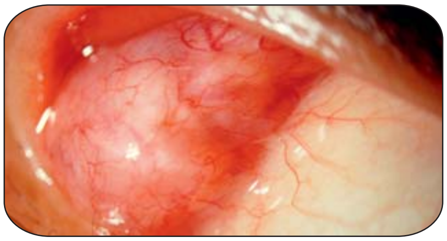
Fig. 2.47B

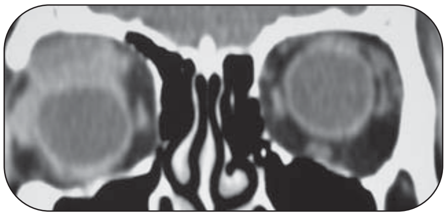
Fig. 2.47C

Lacrimal gland carcinoma

Presentation

- This aggressive tumour presents with a shorter history than an adenoma and is associated with pain.

Signs

- Down and in displacement of the globe.
- Posterior extension may give rise to proptosis (**Fig. 2.48A**), epibulbar congestion, periocular oedema and ophthalmoplegia.
- CT shows a globular mass with an irregular serrated border, often with contiguous erosion or invasion of bone (**Fig. 2.48B**), and occasionally spotty calcification.

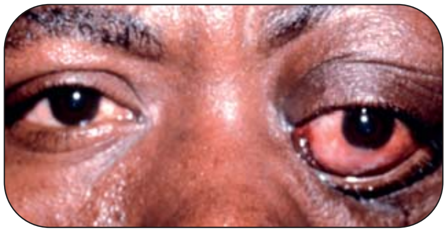
Fig. 2.48A

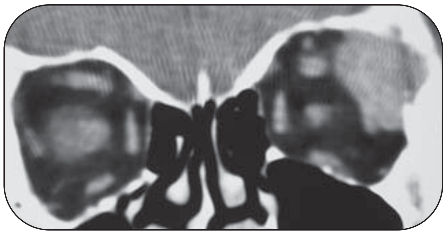
Fig. 2.48B

Dacryops

Definition

- An epithelial cyst caused by obstruction and dilatation of major lacrimal ducts.

Signs

- On eversion of the lid a bluish dome-shaped lesion is apparent (**Fig. 2.49**).

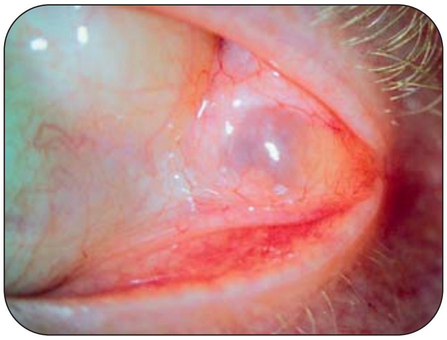

Fig. 2.49

Bilateral

a. *Physiological enlargement* associated with shallow orbits.

b. *Late thyroid eye disease*.

c. *Sarcoidosis* may cause bilateral firm enlarged lacrimal glands.

d. *Lymphomas* typically cause a firm or rubbery enlargement which may be unilateral or bilateral.

e. *Sjögren syndrome* in which bilateral lacrimal gland enlargement is associated with dry eyes and xerostomia.

f. *Mikulicz syndrome* is characterised by hypertrophy of lacrimal and salivary glands associated with dry eyes and xerostomia (**Fig. 2.50**). It may be associated with other diseases such as Sjögren syndrome, sarcoidosis, systemic lupus erythematosus, leukaemia, lymphoma and tuberculosis.

g. *Amyloidosis* may infiltrate the lacrimal glands.

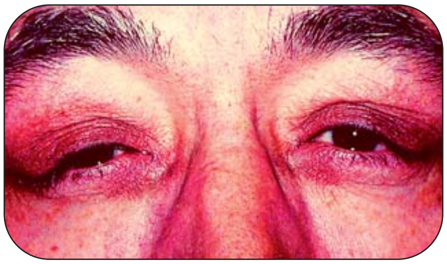

Fig. 2.50

ORBITAL RIM LESIONS

Orbital rim lesions are almost always unilateral and may occur anywhere around the orbital rim. The causes are as follows:

a. *Superficial dermoid cyst* is an asymptomatic firm round lesion, most frequently located in the upper outer quadrant of the orbital margin (**Fig. 2.51**).

b. *Subperiosteal haematoma* is caused by severe trauma.

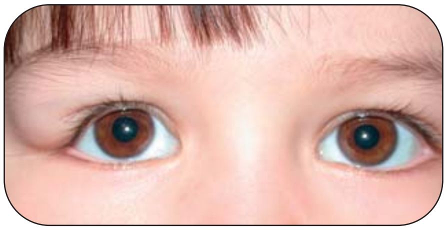

Fig. 2.51

c. *Subperiosteal abscess* is very painful and is associated with orbital cellulitis. It is very dangerous because of the potential for rapid progression and intracranial extension.

DYNAMIC PROPTOSIS

Intermittent

a. *Orbital varices* cause a non-pulsatile proptosis which may be accentuated by the Valsalva manoeuvre (see Fig. 2.38).

b. *Mucocele* may result in proptosis which fluctuates when its walls become inflamed.

c. *Capillary haemangioma* may be associated with increase in proptosis on crying.

d. *Lymphangioma* may cause a proptosis which increases during or after an upper respiratory tract infection.

Pulsatile

a. *Not associated with a bruit* is due to transmitted CSF pulsation. It may be caused by encephalocele in which the pulsation is transmitted to the orbit through a defect in the sphenoid bone, or an orbital roof fracture.

b. *Associated with a bruit* due to arterial pulsation. It may be caused by carotid–cavernous fistula or a congenital arteriovenous communication which may occur in isolation or as part of Wyburn-Mason syndrome.

Conjunctiva 3

CONJUNCTIVITIS 63

Signs of conjunctival inflammation 63
- *Conjunctival injection* 63
- *Papillary reaction* 63
- *Follicular reaction* 63
- *Discharge* 64
- *Membranes* 64
- *Haemorrhages* 64
- *Preauricular lymphadenopathy* 64

Acute papillary conjunctivitis 65
- *Simple bacterial* 65
- *Gonococcal* 65
- *Allergic rhinoconjunctivitis* 65

Acute follicular conjunctivitis 66
- *Adenoviral* 66
- *Herpes simplex* 66
- *Other viral causes* 66

Chronic papillary conjunctivitis 67
- *Vernal keratoconjunctivitis* 67
- *Atopic keratoconjunctivitis* 67
- *Giant papillary conjunctivitis* 68
- *Superior limbic keratoconjunctivitis* 68

Chronic follicular conjunctivitis 69
- *Adult inclusion (TRIC)* 69
- *Trachoma* 69
- *Molluscum contagiosum* 69
- *Chronic toxic* 70
- *Parinaud oculoglandular syndrome* 70

Neonatal conjunctivitis 71
- *Chemical* 71
- *Gonococcal* 71
- *Chlamydial* 71
- *Simple bacterial* 71
- *Herpes simplex type 2* 71
- *Congenital dacryostenosis* 72

Cicatrising conjunctivitis in blistering dermatoses 72
- *Signs of cicatrising conjunctivitis* 72
- *Mucous membrane pemphigoid* 72
- *Bullous pemphigoid* 73
- *Stevens–Johnson syndrome* 73
- *Toxic epidermal necrolysis (Lyell disease)* 73
- *Epidermolysis bullosa* 73
- *Dermatitis herpetiformis* 74
- *Pemphigus vulgaris* 74

Conjunctivitis in systemic disease 74
- *Reiter syndrome* 74
- *Thyrotoxicosis* 75
- *Kawasaki disease* 75
- *Cat-scratch fever* 75
- *Scleroderma* 76

Secondary chronic 'conjunctivitis' 76
- *Chronic blepharitis* 76
- *Keratoconjunctivitis sicca* 76
- *Acne rosacea* 77
- *Chronic canaliculitis* 77
- *Chronic dacryocystitis* 77
- *Floppy eyelid syndrome* 77
- *Mucus fishing syndrome* 78
- *Masquerade syndrome* 78

VASCULAR DILATATION AND MALFORMATION 79
- *Sentinel vessels* 79
- *Carotid–cavernous fistula* 79
- *Varices* 79
- *Active thyroid ophthalmopathy* 79
- *Haemorrhagic lymphangiectasia* 80
- *Sturge–Weber syndrome* 80
- *Louis-Bar syndrome (ataxia-telangiectasia)* 80
- *Fabry disease* 81
- *Rendu–Osler–Weber disease (hereditary haemorrhagic telangiectasia)* 81
- *Radiation telangiectasia* 81

PIGMENTED LESIONS 82

Flat 82
- *Freckle* 82
- *Benign epithelial (racial) melanosis* 82
- *Primary acquired melanosis* 82
- *Congenital melanocytosis* 82
- *Adrenochrome deposits* 83
- *Argyrosis* 83

Elevated 83
- *Naevus* 83
- *Primary melanoma* 84
- *Ciliary body melanoma with extraocular extension* 84
- *Melanocytoma* 84

NON-PIGMENTED FOCAL LESIONS 85

Nodular 85
- *Naevus* 85
- *Primary melanoma* 85
- *Squamous papilloma* 85
- *Pyogenic granuloma* 85
- *Epibulbar dermoid* 86
- *Conjunctival intraepithelial hyperplasia* 86
- *Invasive squamous cell carcinoma* 87

Plaque-like 87
- *Pterygium* 87
- *Pseudo-pterygium* 87
- *Lipodermoid* 87
- *Orbital fat herniation* 88
- *Non-Hodgkin lymphoma and reactive lymphoid hyperplasia* 88
- *Amyloidosis* 88
- *Reactive pseudo-epitheliomatous hyperplasia* 88
- *Benign hereditary intraepithelial dyskeratosis* 89

Cystic 89
- *Primary retention cyst* 89
- *Secondary inclusion cyst* 89

CONJUNCTIVITIS

Signs of conjunctival inflammation

Conjunctival injection

The conjunctival vessels are most superficial and thin, and apart from the major arteries and veins, are virtually invisible to the naked eye.

Signs

- In conjunctivitis the injection is diffuse, beefy-red and is least at the limbus and maximal towards the fornices (**Fig. 3.1**).

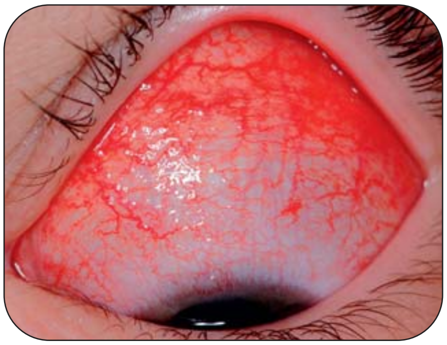

Fig. 3.1

Papillary reaction

Papillae are hyperaemic flat-topped elevations separated by pale outlines which occur on the palpebral tarsal conjunctiva and occasionally on the perilimbal conjunctiva. The clinical appearance of papillae varies greatly according to size and number, and can be arbitrarily graded 0–4.

- *Grade 1* – micropapillae are tiny slightly elevated red dots that give rise to a smooth velvety appearance (**Fig. 3.2**).
- *Grade 2* – macropapillae are less than 1 mm in diameter (**Fig. 3.3**).
- *Grade 3* – giant papillae are more than 1 mm in diameter (**Fig. 3.4**).
- *Grade 4* – enormous and protruding (**Fig. 3.5**).

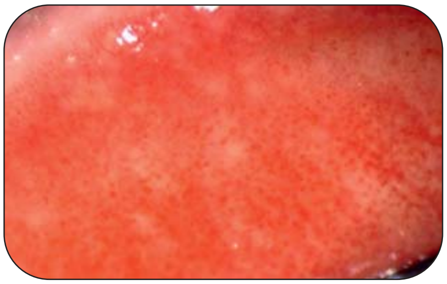

Fig. 3.2

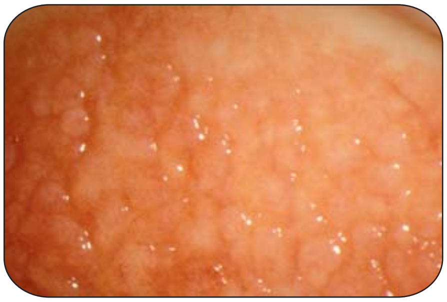

Fig. 3.3

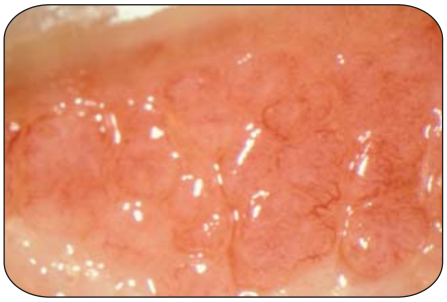

Fig. 3.4

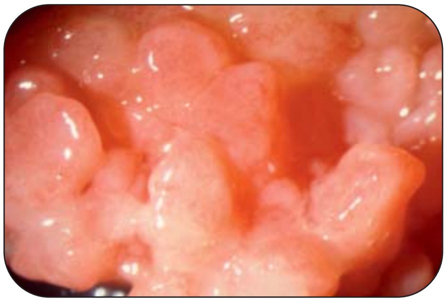

Fig. 3.5

Follicular reaction

Signs

- Discrete, yellowish, slightly elevated lesions most prominent in the fornices (**Fig. 3.6**).
- Blood vessels normally pass over the surface of a follicle and then as it enlarges the vessels are displaced peripherally.

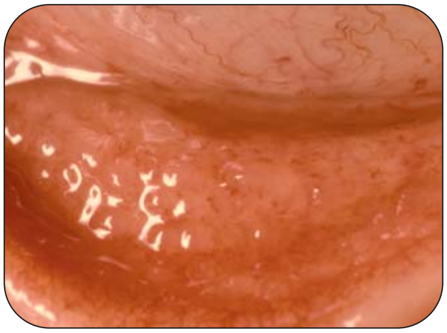

Fig. 3.6

Discharge

a. *Watery* occurs in acute viral and allergic disease.

b. *Mucoid* is typical of chronic allergic disease and dry eye.

c. *Mucopurulent* occurs in acute bacterial and chlamydial conjunctivitis.

d. *Purulent* is typical of gonococcal infection.

Membranes

Signs

- Coagulated exudate adherent to inflamed conjunctival epithelium (**Fig. 3.7**).
- Clinically, a true membrane causes bleeding on attempted removal and a pseudo-membrane does not, but this rule is not infallible.

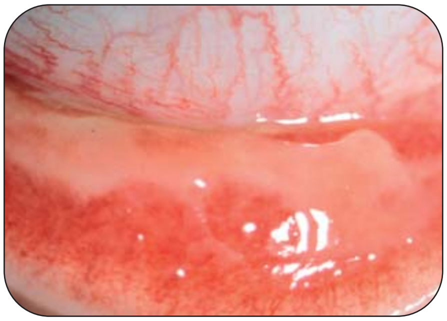
Fig. 3.7

Causes

The causes of true membranes and pseudo-membranes are essentially similar:

- Severe adenoviral conjunctivitis.
- Gonococcal conjunctivitis.
- Ligneous conjunctivitis.
- Acute Stevens–Johnson syndrome.
- Bacterial infection with *Streptococcus* spp. and *Corynebacterium diphtheriae*.

Haemorrhages

Causes

Adenoviral infection and occasionally bacterial infections caused by *S. pneumoniae*, *H. influenzae* and *N. meningitis.*

Preauricular lymphadenopathy (**Fig. 3.8**)

Causes

- Viral infection.
- Chlamydial infection.
- Gonococcal infection.
- Parinaud oculoglandular syndrome.

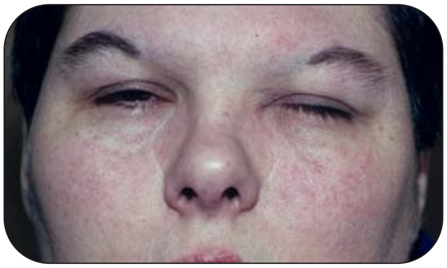
Fig. 3.8

Acute papillary conjunctivitis

Simple bacterial

Signs

- *Eyelids* – crusty and covered by exudate (**Fig. 3.9**).
- *Papillae* – grade 1 (bilateral).
- *Discharge* – initially catarrhal and later mucopurulent (**Fig. 3.10**).
- *Membranes* – usually absent.
- *Cornea* – normal.
- *Preauricular adenopathy* – absent.

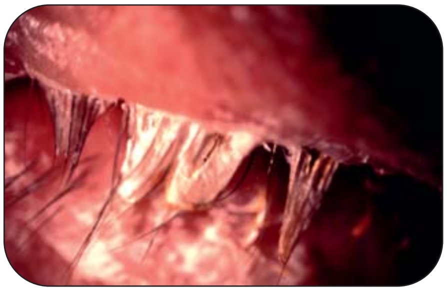

Fig. 3.9

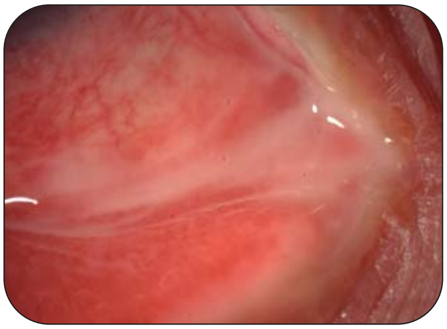

Fig. 3.10

Gonococcal

Signs

- *Eyelids* – oedematous.
- *Papillae* – grade 1 (bilateral).
- *Discharge* – copious purulent.
- *Membranes* – present.
- *Cornea* – ulceration if treatment is delayed (**Fig. 3.11**).
- *Preauricular adenopathy* – present.

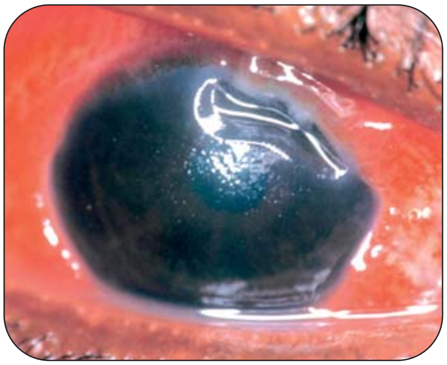

Fig. 3.11

Look for

- In males – urethral discharge and dysuria.
- In females – vaginal discharge and less frequently dysuria.

Allergic rhinoconjunctivitis

Signs

- *Eyelids* – oedematous.
- *Papillae* – grade 1 (bilateral) and often associated with chemosis (**Fig. 3.12**).
- *Discharge* – watery.
- *Membranes* – absent.
- *Cornea* – normal.
- *Preauricular adenopathy* – absent.

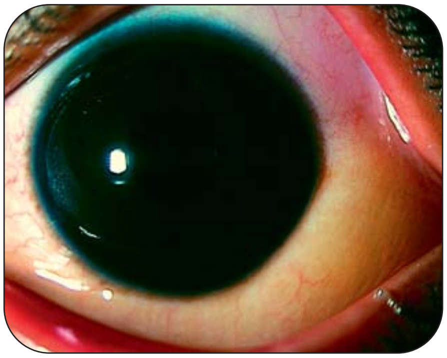

Fig. 3.12

Acute follicular conjunctivitis

Adenoviral

Signs

- *Eyelids* – oedematous.
- *Follicles* – (bilateral) most apparent in the inferior fornices.
- *Discharge* – watery.
- *Membranes* – if severe.
- *Subconjunctival haemorrhages* – if severe.
- *Cornea* – punctate epithelial keratitis is common. It may be followed by subepithelial opacities and anterior stromal infiltrates (**Fig. 3.13**).
- *Preauricular adenopathy* – may be present.

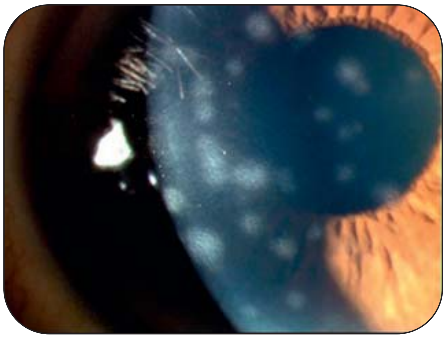
Fig. 3.13

Herpes simplex

Signs

- *Eyelids* – vesicles.
- *Follicles* – (unilateral) most apparent in the inferior fornix.
- *Discharge* – watery.
- *Membranes* – absent.
- *Subconjunctival haemorrhages* – absent.
- *Cornea* – small dendritic ulcers are uncommon (**Fig. 3.14**).
- *Preauricular adenopathy* – present.

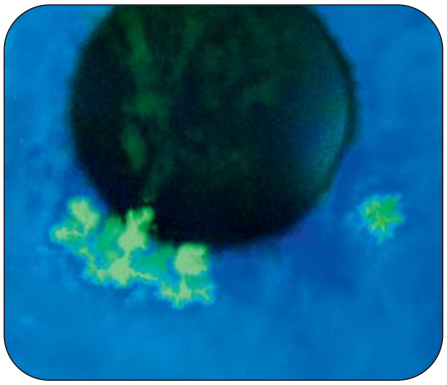
Fig. 3.14

Other viral causes

a. *Epstein–Barr.*

b. *Measles.*

c. *Mumps.*

d. *Newcastle disease.*

e. *Rubella.*

f. *Picornaviruses.*

Chronic papillary conjunctivitis

Vernal keratoconjunctivitis

Definition

- A common recurrent bilateral condition which affects children and young adults.

Signs

- *Eyelids* – normal or may show eczema.
- *Discharge* – mucoid.
- *Papillae* – on superior tarsus grade 2–3, rarely 4.
- *Limbitis* – grade 3 papillae associated with small white (Trantas) dots (**Fig. 3.15**).
- *Conjunctival scarring* – absent.
- *Cornea* – superior corneal punctate epithelial erosions, epithelial macroerosions, shield ulcers and plaques (**Fig. 3.16**), pseudogerontoxon and peripheral vascularisation.
- *Preauricular adenopathy* – absent.

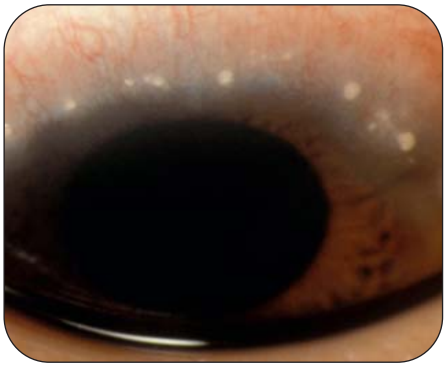

Fig. 3.15

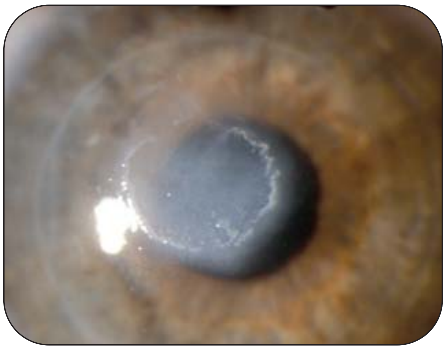

Fig. 3.16

Atopic keratoconjunctivitis

Definition

- An uncommon, serious, bilateral condition which occurs in adults with atopic dermatitis.

Signs

- *Eyelids* – eczema (see Fig. 1.26), madarosis and staphylococcal blepharitis.
- *Discharge* – mucoid.
- *Papillae* – grade 1–2 most intense on the inferior palpebral conjunctiva.
- *Limbitis* – absent.
- *Conjunctival scarring* – featureless appearance of the superior tarsal conjunctiva (**Fig. 3.17**).
- *Cornea* – inferior punctate epithelial erosions, persistent epithelial defects (**Fig. 3.18**) and peripheral vascularisation.
- *Preauricular adenopathy* – absent.

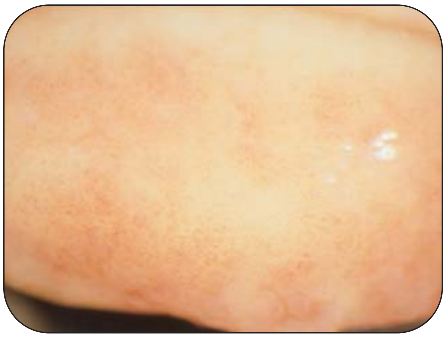

Fig. 3.17

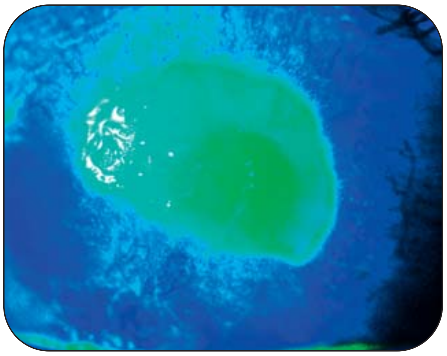

Fig. 3.18

Table 3.1 **Vernal versus atopic disease**

	Vernal	Atopic
Age	Younger	Older
Gender predilection	Males	Nil
Duration of disease	Self-limiting	Chronic
Time of year	Spring	Perennial
Conjunctival involvement	Superior tarsus	Inferior tarsus
Cicatrisation	Absent	Common
Cornea	Shield ulcer	Persistent epithelial defect
Corneal scarring	Mild	Severe
Corneal vascularisation	Rare	Common

Giant papillary conjunctivitis

Definition

- A unilateral or bilateral condition, depending on the cause such as inappropriate contact lens wear, particularly soft, ocular prosthetics (**Fig. 3.19**) and protruding sutures following surgery.

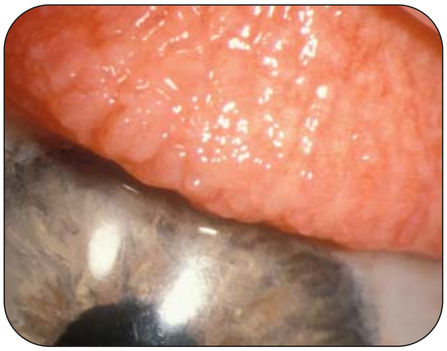

Fig. 3.19

Signs

- *Eyelids* – normal.
- *Discharge* – mucoid.
- *Papillae* – grade 2–3.
- *Limbitis* – absent.
- *Cornea* – normal.
- *Preauricular adenopathy* – absent.

Superior limbic keratoconjunctivitis

Definition

- An uncommon, usually bilateral condition which is frequently associated with thyroid dysfunction.

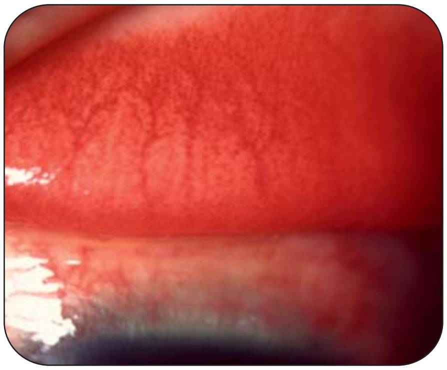

Fig. 3.20

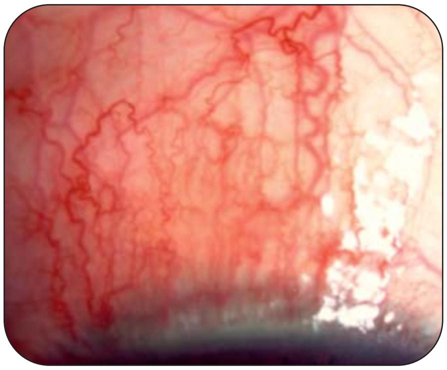

Fig. 3.21

Signs

- *Eyelids* – normal or may show lid lag and retraction if associated with thyroid eye disease.
- *Discharge* – mucoid.
- *Papillae* – grade 1 on superior tarsal conjunctiva (**Fig. 3.20**).
- *Superior conjunctiva* – hyperaemic and thickened (**Fig. 3.21**).
- *Limbitis* – mild.
- *Cornea* – superior punctate erosions and filaments.
- *Preauricular adenopathy* – absent.

Chronic follicular conjunctivitis

Adult inclusion (TRIC)

Definition

- Usually a unilateral condition caused by *C. trachomatis* serotypes D–K.

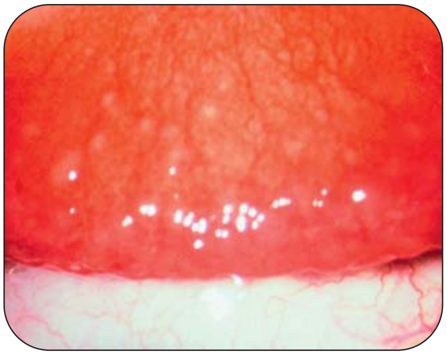

Fig. 3.22

Signs

- *Eyelids* – normal.
- *Discharge* – mucopurulent.
- *Follicles* – large in the inferior fornices and less prominent in the superior tarsal conjunctiva (**Fig. 3.22**).
- *Limbitis* – occasionally limbal follicles.
- *Cornea* – peripheral infiltrates (see Fig. 5.121).
- *Preauricular adenopathy* – present.

Look for

- In males – ‘non-specific urethritis’.
- In females – abacterial pyuria and cervicitis.

Trachoma

Definition

- A bilateral condition caused by *C. trachomatis* serotypes A, B, Ba and C that is associated with poverty and poor hygiene.

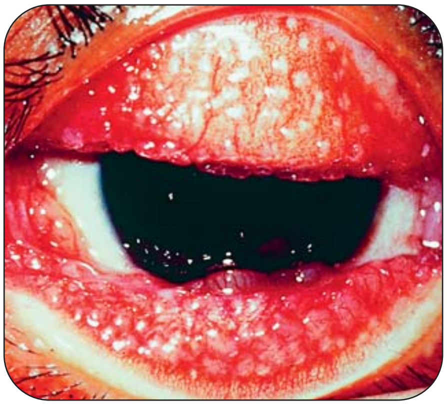

Fig. 3.23

Signs

- *Eyelids* – scarring, trichiasis and cicatricial entropion in advanced disease.
- *Discharge* – mucopurulent.
- *Follicles* – initially mixed with papillae (**Fig. 3.23**).
- *Limbitis* – may give rise to a row of shallow depressions (Herbert pits) following resolution (**Fig. 3.24**).
- *Cornea* – scarring.
- *Preauricular adenopathy* – absent.

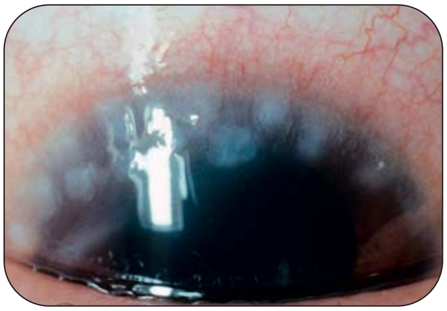

Fig. 3.24

Molluscum contagiosum

Definition

- An uncommon unilateral condition caused by a poxvirus.

Signs

- *Eyelids* – umbilicated nodule on the lid margin (**Fig. 3.25**).
- *Discharge* – mild mucoid.
- *Follicles* – in inferior fornix and occasionally on the bulbar conjunctiva.
- *Cornea* – epithelial keratitis or micropannus in chronic cases.
- *Preauricular adenopathy* – absent.

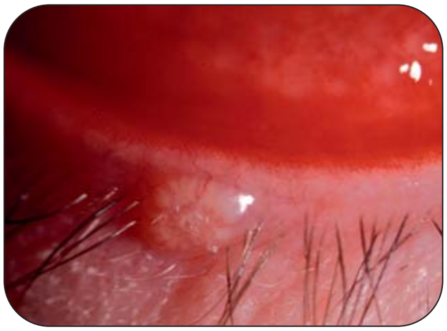

Fig. 3.25

Chronic toxic

Causes

- Topical eye medication.
- Eye make-up.
- Variety of environmental pollutants.

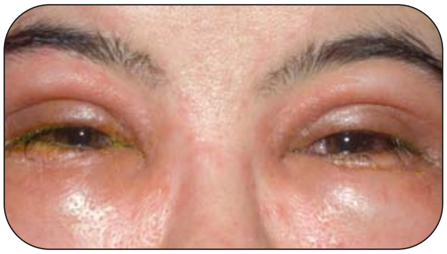

Fig. 3.26

Signs

- *Eyelids* – may show signs of contact dermatitis (**Fig. 3.26**).
- *Discharge* – watery.
- *Follicles* – initially the reaction is papillary; follicles develop after several weeks and are most prominent in the inferior fornix and inferior palpebral conjunctiva.
- *Cornea* – inferior punctate erosions.
- *Preauricular adenopathy* – absent.

Parinaud oculoglandular syndrome

Definition

- A rare usually unilateral condition that has many diverse causes (see below).

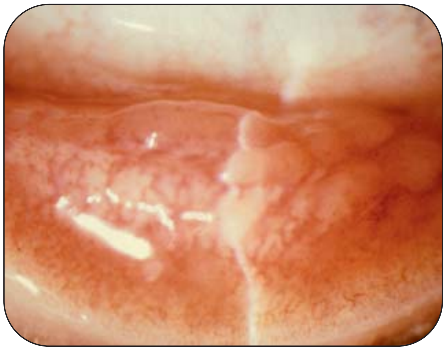

Fig. 3.27

Signs

- *Eyelids* – normal.
- *Discharge* – mucoid.
- *Conjunctiva* – ulceration or large follicles associated with granulomas (**Fig. 3.27**).
- *Cornea* – normal.
- *Cervical and preauricular adenopathy* – severe.

Causes

- Common – cat-scratch disease, tularaemia and sporotrichosis.
- Occasional – TB, syphilis and coccidioidomycosis.
- Rare – sarcoidosis, lymphogranuloma venereum and chancroid.

Neonatal conjunctivitis

Neonatal conjunctivitis is defined as conjunctival inflammation occurring during the first month of life.

Chemical

Presentation

- Within the first few days of life.

Cause

- Topical administration of silver nitrate at the time of birth.

Signs

- Mild diffuse conjunctival injection without discharge.

Gonococcal

Presentation

- Within 1–3 days of life.

Signs

- Copious purulent discharge (**Fig. 3.28**) which may be associated with membrane formations.

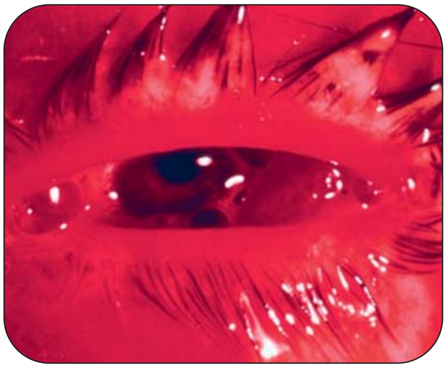

Fig. 3.28

Chlamydial

Presentation

- Within 5–25 days but the interval may be longer if erythromycin has been used prophylactically.

Signs

- Initially unilateral and then bilateral papillary conjunctivitis (infant cannot produce follicles).

Simple bacterial

This may occur at any time.

Herpes simplex type 2

This is rare and may be associated with lid vesicles.

Congenital dacryostenosis

Presentation

- Soon after birth with usually unilateral discharge and epiphora (**Fig. 3.29**) with reflux of mucopurulent material on massaging the lacrimal sac.

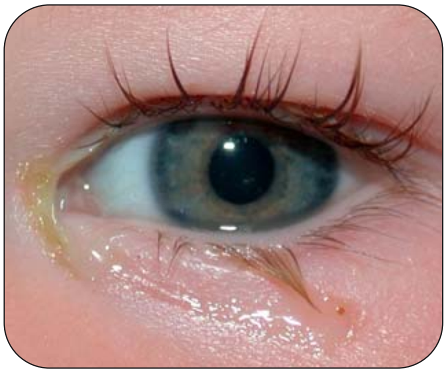

Fig. 3.29

Cicatrising conjunctivitis in blistering dermatoses

Signs of cicatrising conjunctivitis

- Progressive subconjunctival fibrosis and obliteration of the fornices.
- Flattening of the plica and keratinisation of the caruncle (**Fig. 3.30**).
- Symblepharon formation (**Fig. 3.31**).
- Keratopathy resulting from drying, exposure and keratinisation (**Fig. 3.32**).

The following are associated blistering dermatoses.

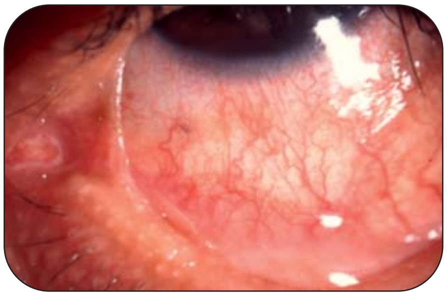

Fig. 3.30

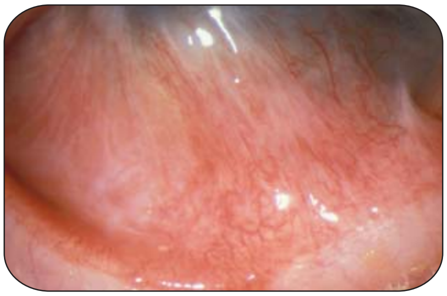

Fig. 3.31

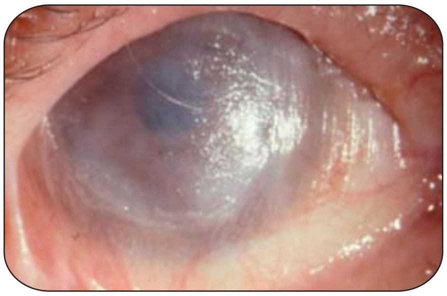

Fig. 3.32

Mucous membrane pemphigoid

Conjunctivitis

- Affects the majority of cases.

Signs

- Oral blisters are very common (**Fig. 3.33** and see Fig. 15.30).
- Skin blisters are sparse.

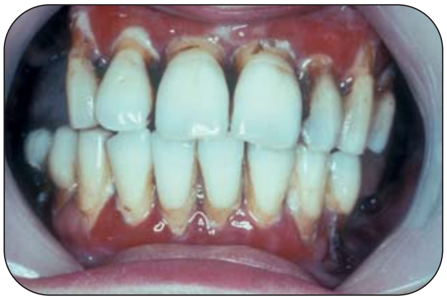

Fig. 3.33

Bullous pemphigoid

Conjunctivitis

- Is uncommon.

Signs

- Oral blisters are rare.
- Skin blisters are large, tense and often on the limbs and trunk (**Fig. 3.34**).

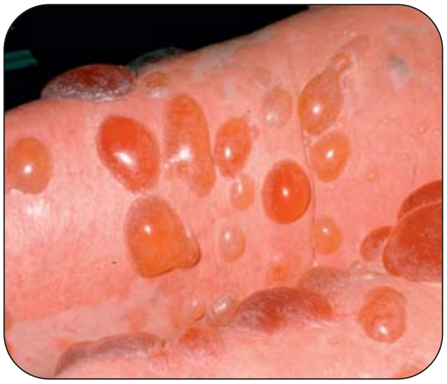

Fig. 3.34

Stevens–Johnson syndrome

Pathogenesis

- Triggered by drugs or infections.

Conjunctivitis

- Is common and usually self-limiting although cicatrisation may occur in some cases.

Signs

- Oral bullae with haemorrhagic crusting of the lips are characteristic (see Fig. 15.29).
- Target skin lesions involving the palms (**Fig. 3.35**) and soles are common but blisters are uncommon.

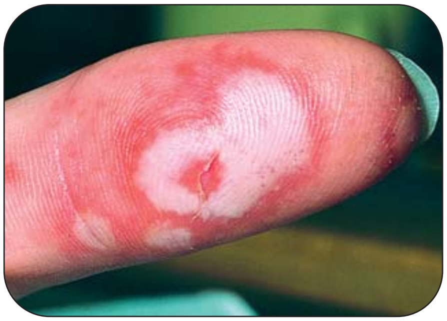

Fig. 3.35

Toxic epidermal necrolysis (Lyell disease)

Conjunctivitis

- Is common but milder than in Stevens–Johnson syndrome.

Signs

- Skin blisters are painful, transient and widespread, and resemble scalded skin (**Fig. 3.36**).

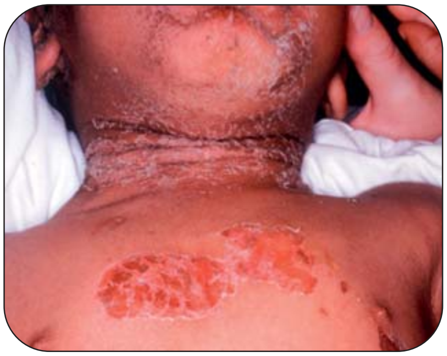

Fig. 3.36

Epidermolysis bullosa

Conjunctivitis

- Is common.

Signs

- Skin blistering induced by minimal trauma (**Fig. 3.37**).
- Nail dystrophy, mitten-hands and feet with partial syndactyly are common.

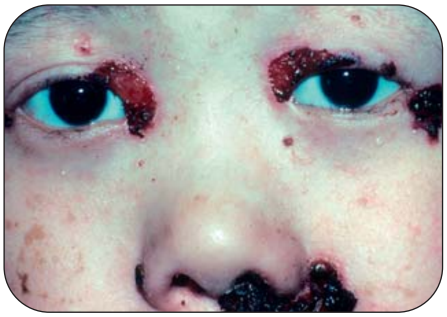

Fig. 3.37

Dermatitis herpetiformis

Conjunctivitis

- Is uncommon.

Signs

- Skin blisters are small, itchy and occur symmetrically in groups on extensor surfaces (**Fig. 3.38**).

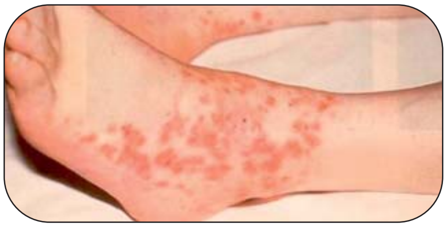

Fig. 3.38

Pemphigus vulgaris

Conjunctivitis

- Is uncommon.

Signs

- Oral blisters are common and are often the presenting feature (see Fig. 15.31).
- Flaccid, superficial skin blisters erode and leave weeping areas (**Fig. 3.39**).

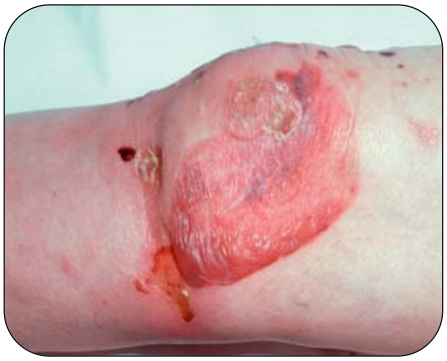

Fig. 3.39

Table 3.2 **Cicatricial pemphigoid vs Stevens–Johnson syndrome**

Feature	Cicatricial pemphigoid	Stevens–Johnson
Aetiology	Autoimmune	Reaction to drugs and micro-organisms
Age	Elderly	Any age
Onset	Insidious	Acute
Course	Limited	Chronic-remittent
Oral involvement	Yes	Yes
Genital involvement	Yes	Yes
Skin blisters	In severe disease	Uncommon and sparse

Conjunctivitis in systemic disease

Reiter syndrome

Definition

- A spondyloarthropathy that typically affects young men.

Conjunctivitis

- Acute, bilateral, mucopurulent and self-limiting.

Look for

- Acute anterior uveitis.
- Urethritis.
- Asymmetric arthritis most frequently involving the knees and ankles.
- Circinate balanitis (see Fig. 15.102).
- Keratoderma blenorrhagica involving the palms and soles (**Fig. 3.40**).

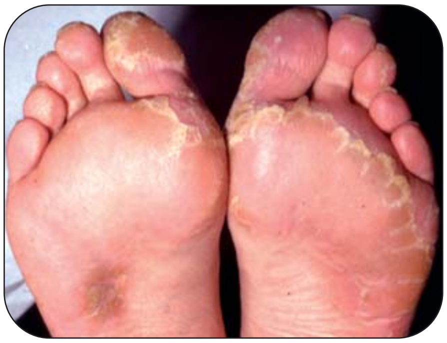

Fig. 3.40

Thyrotoxicosis

Signs

- Bilateral asymmetrical superior limbic keratoconjunctivitis (see Figs 3.20 and 3.21).

Look for

- Thyroid enlargement (**Fig. 3.41**).
- Fine tremor of the hands.
- Tachycardia and atrial fibrillation.

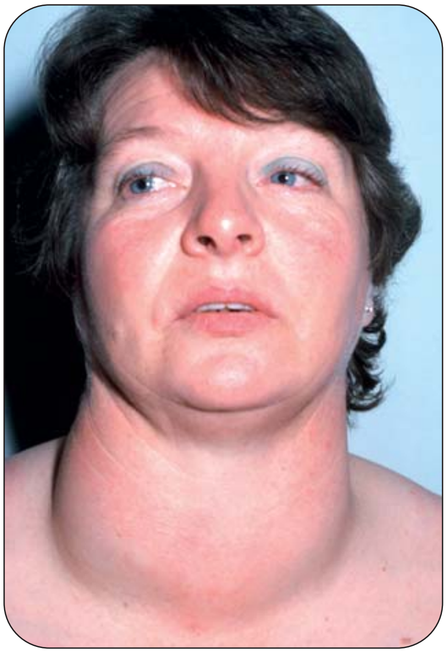

Fig. 3.41

Kawasaki disease

Definition

- A rare idiopathic vasculitis-like disease affecting children.

Conjunctivitis

- Bilateral acute conjunctivitis is common.

Look for

- Erythematous rash and cervical lymphadenopathy.
- Cardiac disease and aneurysm formation.
- Oedema of the extremities (**Fig. 3.42**).

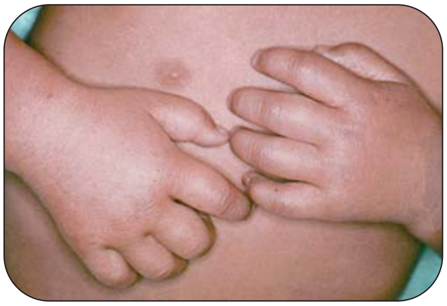

Fig. 3.42

Cat-scratch fever

Cause

- Infection with *Bartonella henselae* transmitted by a scratch or bite of an infected cat.

Conjunctivitis

- Parinaud oculoglandular syndrome may occur (see Fig. 3.27).

Look for

- Fever and malaise.
- Skin papules (**Fig. 3.43**) and regional lymphadenopathy.

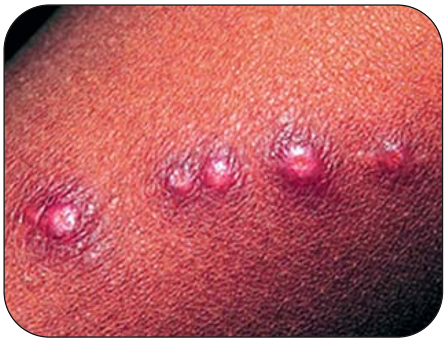

Fig. 3.43

Scleroderma

Conjunctivitis

- Is uncommon and may be associated with mild forniceal shortening.

Look for

- Tight, hard, waxy skin and sclerodactyly (**Fig. 3.44**).
- Raynaud phenomenon (see Fig. 15.60).

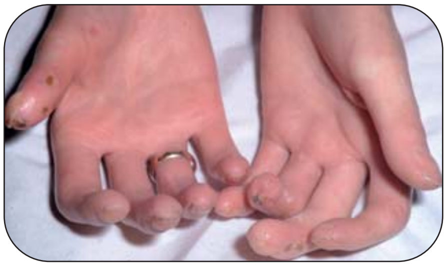

Fig. 3.44

Secondary chronic 'conjunctivitis'

The following conditions should be considered in patients with chronic ocular irritation or discharge without an immediately apparent cause.

Chronic blepharitis

Symptoms

- Chronic non-specific ocular irritation, worse in the mornings.

Look for

- Hard scales and crusts around the lashes (**Fig. 3.45**).
- Atopic and seborrhoeic dermatitis, and acne rosacea.

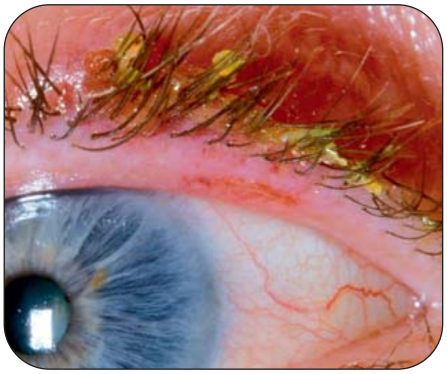

Fig. 3.45

Keratoconjunctivitis sicca

Symptoms

- Bilateral non-specific chronic ocular irritation and mucoid discharge.

Look for

- Reduced or absent tear meniscus.
- Corneal filaments and mucous plaques (see Fig. 5.44).
- Interpalpebral staining of conjunctiva and cornea with rose Bengal (**Fig. 3.46**).
- Reduced tear film break-up time.

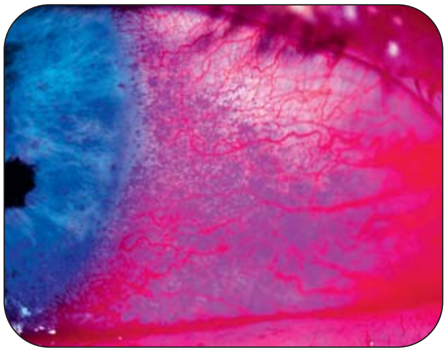

Fig. 3.46

Acne rosacea

Symptoms

- Bilateral, chronic, recurrent non-specific ocular irritation.

Look for

- Posterior blepharitis (see Fig. 1.20).
- Mild conjunctival injection.
- Facial flushing.
- Telangiectasia of the nose, cheeks and forehead (**Fig. 3.47**).

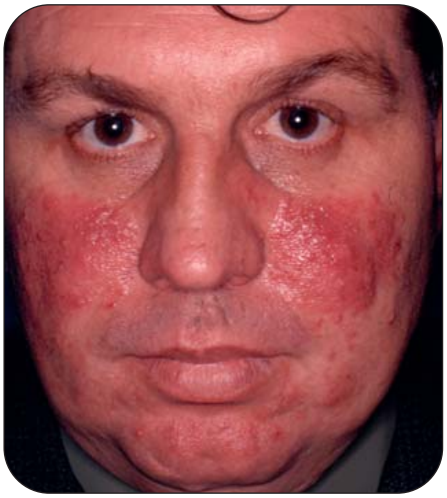

Fig. 3.47

Chronic canaliculitis

Symptoms

- Unilateral, chronic, mucopurulent discharge resistant to topical antibiotics.

Look for

- Pouting punctum and oedema over the canaliculi (**Fig. 3.48**).

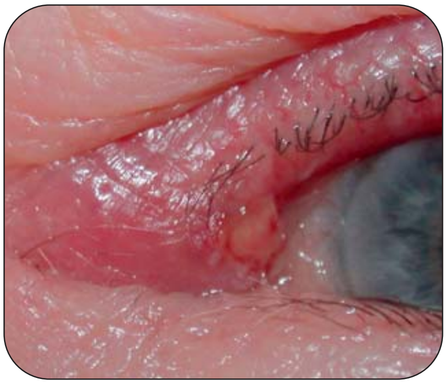

Fig. 3.48

Chronic dacryocystitis

Symptoms

- Unilateral, chronic, mucopurulent discharge and epiphora.

Look for

- Swelling at the medial canthus due to a lacrimal sac mucocele (**Fig. 3.49**).
- Pressure over the sac causes regurgitation of pus and mucus.

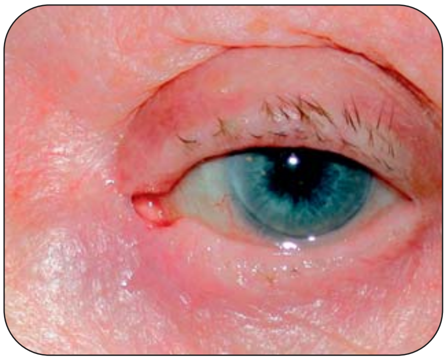

Fig. 3.49

Floppy eyelid syndrome

Definition

- An uncommon unilateral or bilateral condition which typically affects obese men.

Look for

- Loose upper lid skin and rubbery easily evertable tarsal plates (**Fig. 3.50**).
- Trauma to the superior tarsal conjunctiva during sleep resulting in chronic ocular irritation.

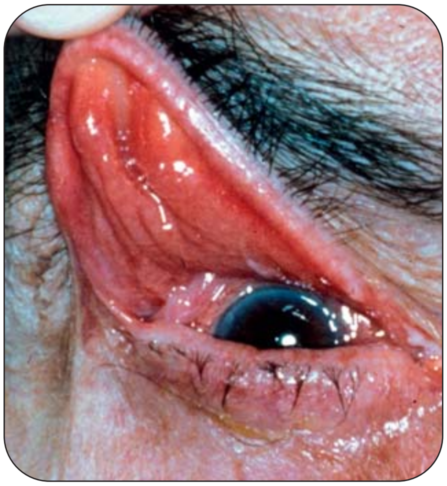

Fig. 3.50

Mucus fishing syndrome

Definition

- A rare, unilateral or bilateral condition, which may be difficult to diagnose because the patient may not admit to self-trauma of the conjunctiva.

Look for

- Conjunctival mucus and isolated areas that stain with rose Bengal (**Fig. 3.51**).

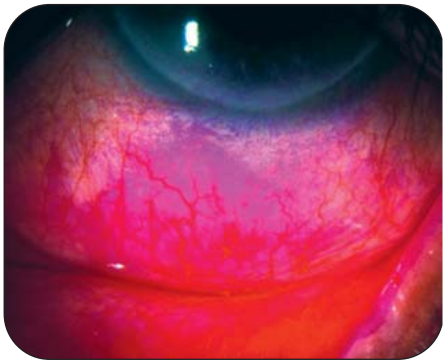

Fig. 3.51

Masquerade syndrome

Signs

- A unilateral condition most frequently caused by infiltration of the conjunctiva by spreading sebaceous gland carcinoma (pagetoid spread – **Fig. 3.52**) which may be mistaken for chronic conjunctival inflammation.

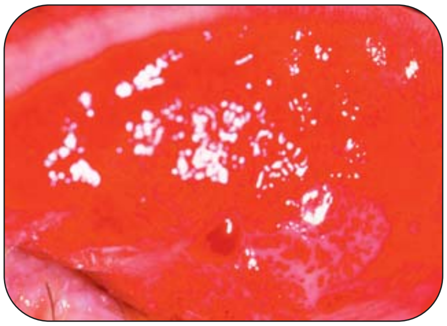

Fig. 3.52

VASCULAR DILATATION AND MALFORMATION

Sentinel vessels

Signs

- Unilateral dilated epibulbar vessel in the same quadrant as an anterior uveal melanoma (**Fig. 3.53**).

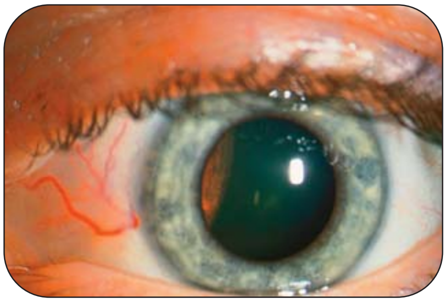

Fig. 3.53

Carotid–cavernous fistula

Signs

- Unilateral dilated episcleral vessels (**Fig. 3.54**).

Look for

- Chemosis.
- Pulsating proptosis with a bruit.
- Elevation of IOP.
- Ophthalmoplegia.
- Retinopathy.

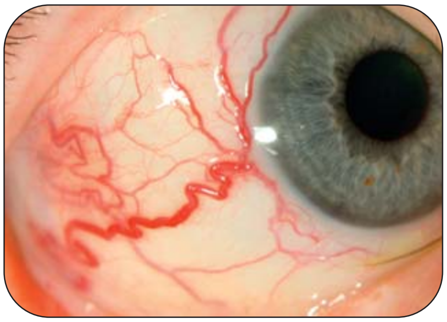

Fig. 3.54

Varices

Signs

- Grape-like vascular dilatations most often in the inferior fornix (**Fig. 3.55**).
- Accentuated on Valsalva manoeuvre.

Look for

- Eyelid (see Fig. 2.40b) and orbital (see Fig. 2.38) involvement.

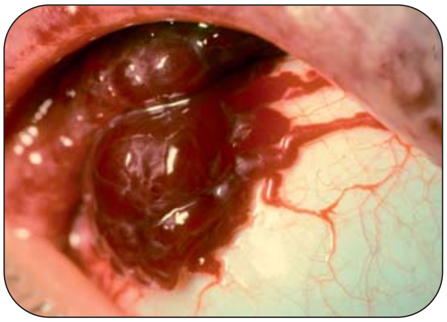

Fig. 3.55

Active thyroid ophthalmopathy

Signs

- Usually bilateral epibulbar hyperaemia overlying a horizontal muscle (**Fig. 3.56**), a sensitive sign of inflammatory activity.

Look for

- Periorbital swelling.
- Chemosis.
- Superior limbic keratoconjunctivitis (see Figs 3.20 and 3.21).

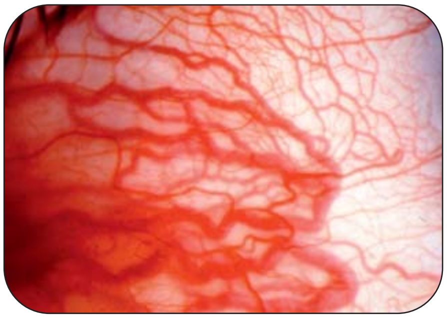

Fig. 3.56

Haemorrhagic lymphangiectasia

Definition

- A rare condition that may occur spontaneously or be precipitated by trauma or inflammation.

Signs

- Irregular sausage-shaped connections between a blood vessel and a lymphatic channel so that the latter is permanently or intermittently filled with blood (**Fig. 3.57**).

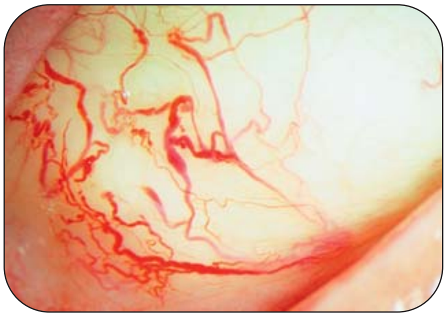
Fig. 3.57

Look for

- Involvement of the lids and orbit.

Sturge–Weber syndrome

Definition

- A sporadic phacomatosis characterised by facial naevus flammeus (port-wine stain – see Fig. 1.48) and ipsilateral leptomeningeal haemangioma.

Signs

- Ipsilateral conjunctival telangiectasia (**Fig. 3.58**) or episcleral haemangioma.

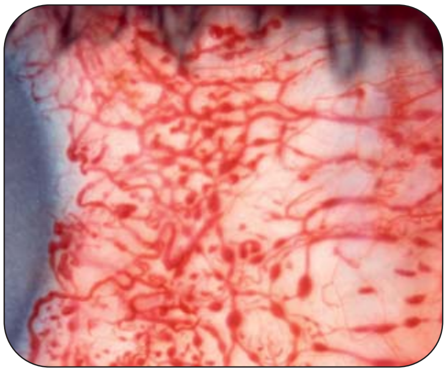
Fig. 3.58

Look for

- Glaucoma.
- Occasional heterochromia iridis.
- Diffuse choroidal haemangioma.

Louis-Bar syndrome (ataxia-telangiectasia)

Signs

- Bulbar conjunctival telangiectasia often with aneurysm formation (**Fig. 3.59**).

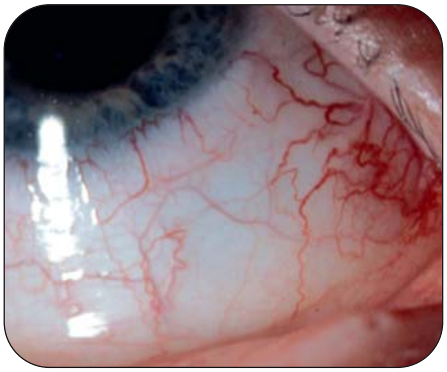
Fig. 3.59

Look for

- Ophthalmoplegia.
- Cerebellar ataxia and nystagmus.
- Cutaneous telangiectasia.
- Lymphopenia, recurrent infections and lymphoma.

Fabry disease

Signs

- Conjunctival telangiectasia (**Fig. 3.60**).

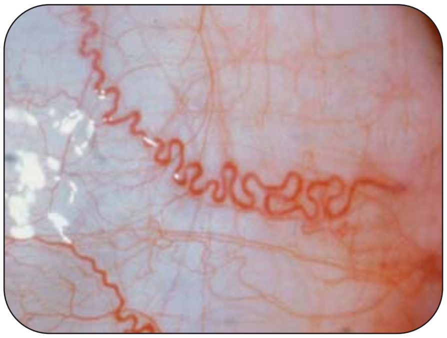

Fig. 3.60

Look for

- Vortex keratopathy.
- Spoke-like cataract.
- Retinal venous tortuosity.
- Third nerve palsy and nystagmus.
- Periodic painful extremities.
- Purple cutaneous telangiectasia (angiokeratoma diffusum – see Fig. 10.34).
- Renal and cardiac disease.

Rendu–Osler–Weber disease (hereditary haemorrhagic telangiectasia)

Signs

- Stellate vascular conjunctival lesions on the palpebral conjunctiva (**Fig. 3.61**).

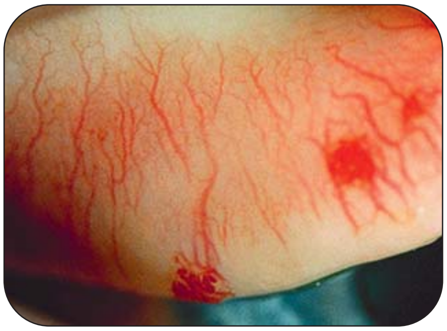

Fig. 3.61

Look for

- Retinal haemorrhages.
- Telangiectasia of skin, lips and mouth.
- Epistaxis and bleeding from gut and lungs.

Radiation telangiectasia

Telangiectasis is an uncommon late complication of radiotherapy of anterior segment neoplasia (**Fig. 3.62**).

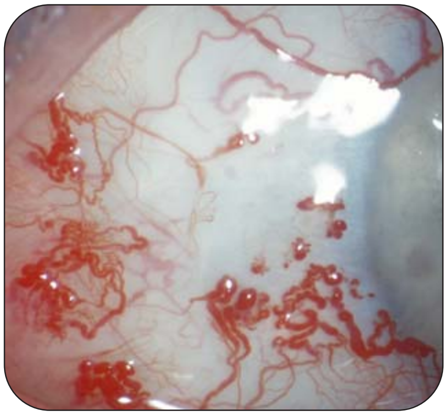

Fig. 3.62

PIGMENTED LESIONS

Flat

Freckle

Signs

- Small isolated spot of pigment (**Fig. 3.63**).

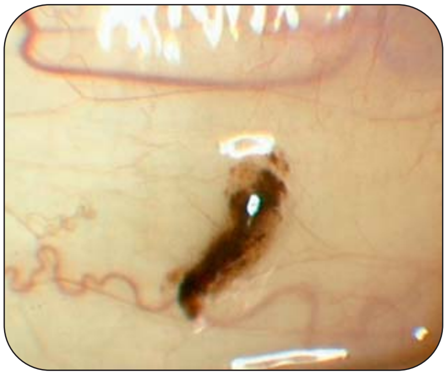

Fig. 3.63

Benign epithelial (racial) melanosis

Signs

- Bilateral, yellow-brown or brown-black, interpalpebral patches most prominent at the limbus that can be moved easily over the globe.
- The pigmentation is often more intense around an intrascleral nerve as it enters the sclera (Axenfeld loop) or the perforating branches of the anterior ciliary vessels (**Fig. 3.64**).

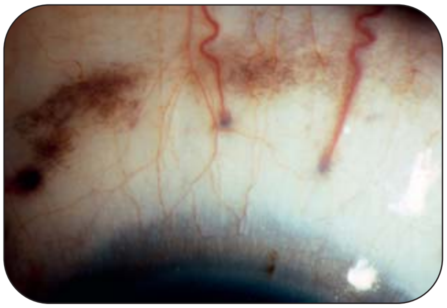

Fig. 3.64

Primary acquired melanosis

Signs

- Dark patches with indistinct margins which may involve any part of the conjunctiva (**Fig. 3.65**).
- Malignant change should be suspected if the patches become nodular.

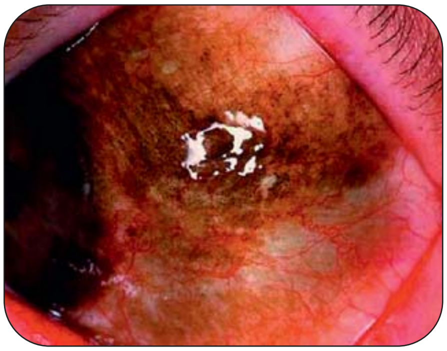

Fig. 3.65

Congenital melanocytosis

Signs

- Unilateral, subepithelial pigmentation that cannot be moved over the globe (**Fig. 3.66**).

Classification

a. *Dermal melanocytosis* involves only skin.

b. *Isolated melanocytosis oculi* involves only the eye.

c. *Oculodermal melanocytosis* (naevus of Ota) involves the skin and eye (see Fig. 1.47).

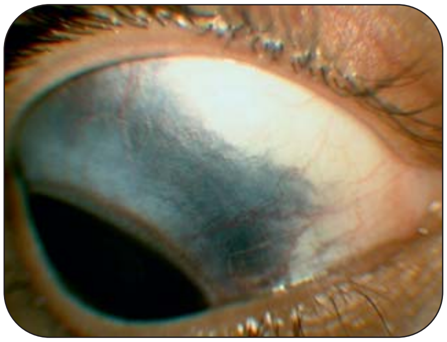

Fig. 3.66

Look for

- Iris hyperpigmentation and mammillations.
- Trabecular hyperpigmentation and increased risk of glaucoma.
- Fundus hyperpigmentation and increased risk of uveal melanoma in Caucasians.

Adrenochrome deposits

Cause

- Long-term use of topical adrenaline (epinephrine) for glaucoma (now seldom, if ever, prescribed).

Signs

- Small discrete clumps of melanin on the tarsal and forniceal conjunctiva (**Fig. 3.67**).

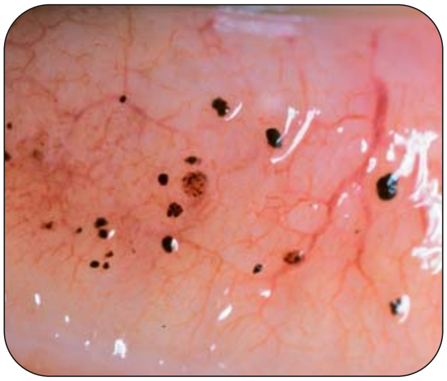

Fig. 3.67

Argyrosis

Cause

- Deposition of silver caused by industrial exposure or topical medication with a silver nitrate compound (now seldom used).

Signs

- Conjunctival pigmentation is diffuse and slate-grey (**Fig. 3.68**).

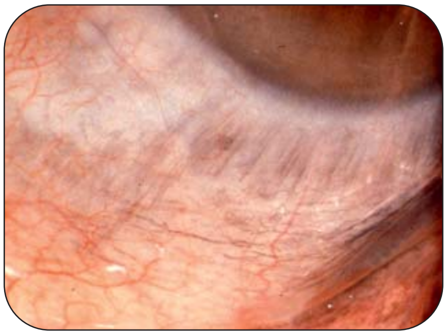

Fig. 3.68

Elevated

Naevus

Signs

- Well-defined, slightly elevated tan or brown lesion which moves freely over the globe and frequently contains cystic spaces.
- Because naevi have a predilection for the limbus (**Fig. 3.69**), plica and caruncle (**Fig. 3.70**), an elevated pigmented lesion elsewhere is unlikely to be a naevus.

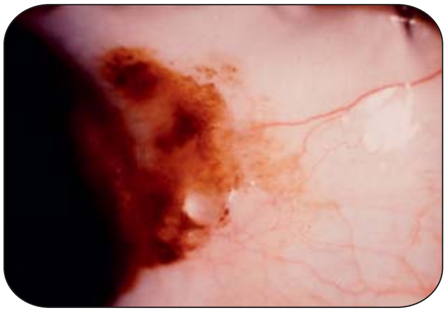

Fig. 3.69

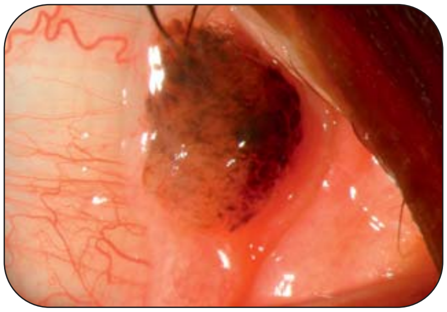

Fig. 3.70

Primary melanoma

Signs

- Pigmented nodule often associated with 'feeder' vessels that has a predilection for the limbus (**Fig. 3.71**) but may affect any part of the conjunctiva.

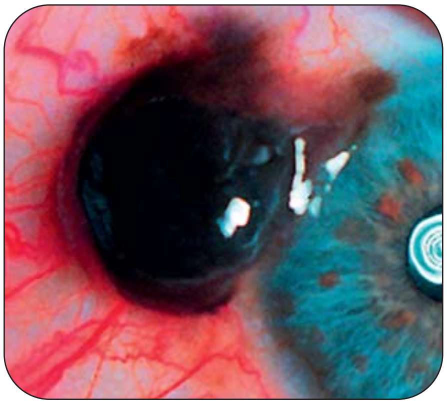

Fig. 3.71

Ciliary body melanoma with extraocular extension

Signs

- Limbal mass which may be mistaken for a primary melanoma on cursory examination (**Fig. 3.72**).

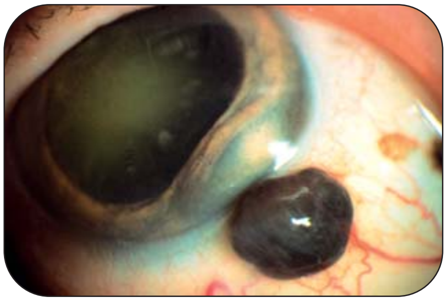

Fig. 3.72

Melanocytoma

Signs

- Black, slightly elevated, slow-growing lesion which does not move freely over the globe (**Fig. 3.73**).

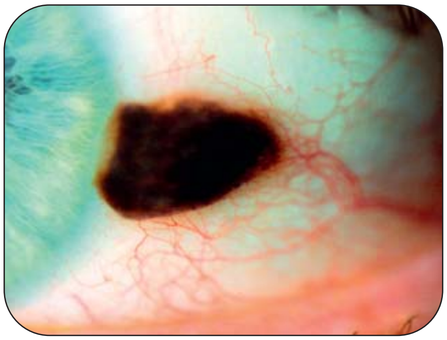

Fig. 3.73

NON-PIGMENTED FOCAL LESIONS

Nodular

Naevus

Signs

- Some naevi may be virtually non-pigmented (**Fig. 3.74**).

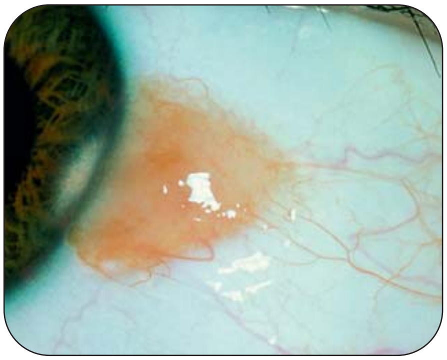

Fig. 3.74

Primary melanoma

Signs

- Nodular lesion with a fish-flesh appearance often associated with ‘feeder’ vessels (**Fig. 3.75**).

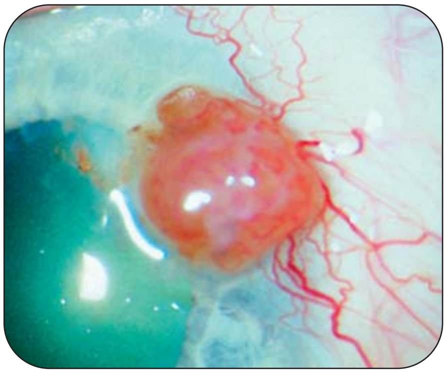

Fig. 3.75

Squamous papilloma

Signs

- Sessile lesions are unilateral and frequently juxtalimbal (**Fig. 3.76**).
- Pedunculated lesions may be bilateral and multiple, and may be located near the limbus, at the caruncle or in the fornices (**Fig. 3.77**).

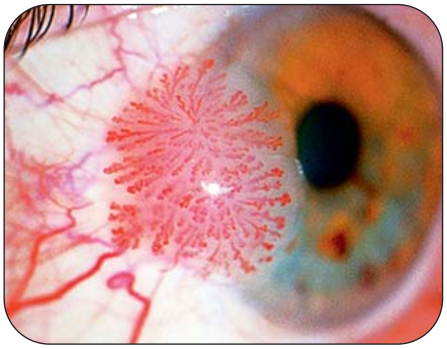

Fig. 3.76

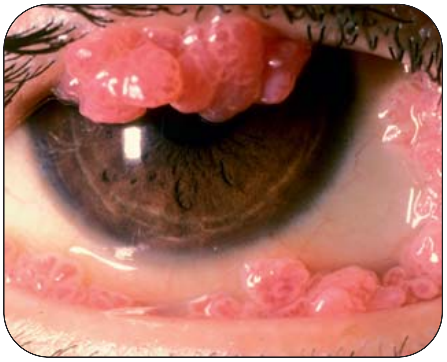

Fig. 3.77

Pyogenic granuloma

Cause

- Conjunctival trauma or surgery.

Signs

- Fast-growing pinkish, fleshy, mass that may have superficial ulceration (**Fig. 3.78**).

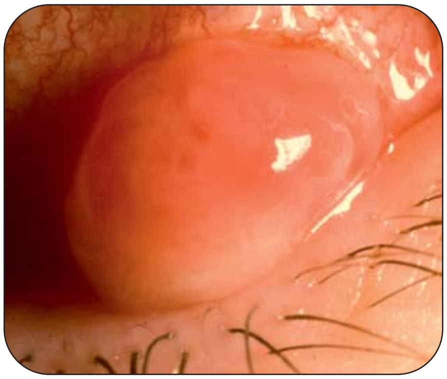

Fig. 3.78

Epibulbar dermoid

Signs

- Unilateral, congenital, smooth, round white mass located at the inferotemporal limbus that may encroach onto the cornea, sometimes with fine emerging hairs (**Fig. 3.79**).

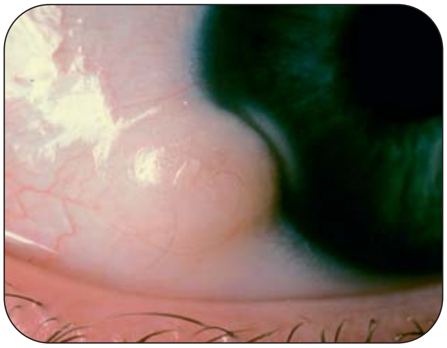

Fig. 3.79

Look for

- Lid coloboma and ocular coloboma.
- Microphthalmos.
- Aniridia.
- Goldenhar syndrome.
- Treacher Collins syndrome.

Conjunctival intraepithelial hyperplasia

Definition

- A rare, unilateral, slow-growing juxtalimbal condition that may spread to adjacent cornea.

Signs

- Gelatinous lesion with superficial vessels (**Fig. 3.80**).
- Less frequently the lesion is covered by a white leukoplakic plaque (**Fig. 3.81**).
- Papillary configuration is the least common (**Fig. 3.82**).

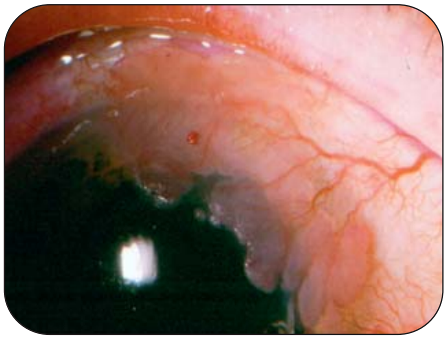

Fig. 3.80

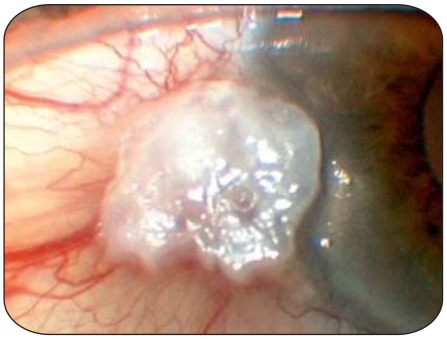

Fig. 3.81

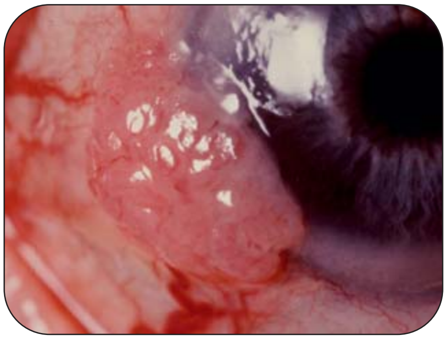

Fig. 3.82

Invasive squamous cell carcinoma

Definition

- Slow-growing, locally invasive tumour which occurs most frequently at the limbus.

Signs

- Although similar to conjunctival intraepithelial hyperplasia the tumour is usually more elevated, involves a greater area of the limbus and is associated with prominent feeder vessels (**Fig. 3.83**).

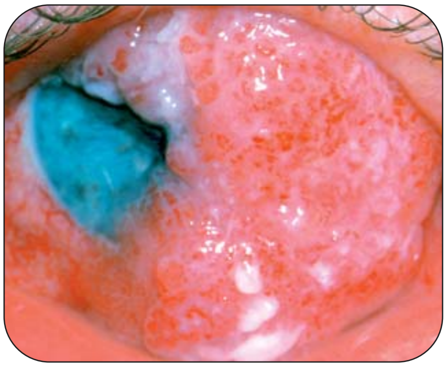

Fig. 3.83

Plaque-like

Pterygium

Signs

- Triangular, fibrovascular, subconjunctival lesion that extends onto the nasal cornea and is fixed to underlying structures throughout (**Fig. 3.84**).

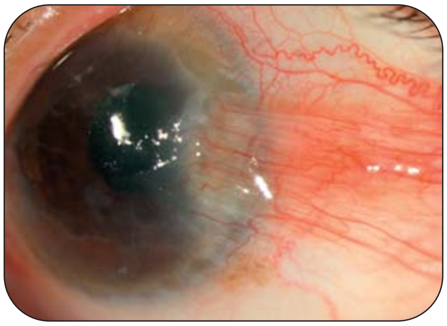

Fig. 3.84

Pseudo-pterygium

Cause

- Adhesion of a fold of conjunctiva to a peripheral corneal ulcer or an area of thinning.

Signs

- Similar to a pterygium but not necessarily located nasally and is fixed to the cornea only at its apex (**Fig. 3.85**).

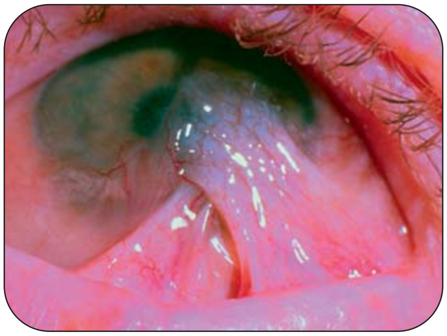

Fig. 3.85

Lipodermoid

Signs

- Large, usually bilateral, movable, subconjunctival lesion located at the outer canthus (**Fig. 3.86**).

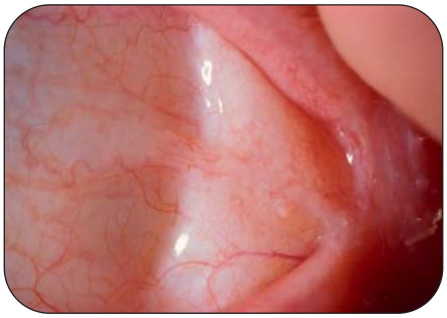

Fig. 3.86

Orbital fat herniation

Signs

- Similar to a lipodermoid but more mobile and softer (**Fig. 3.87**).

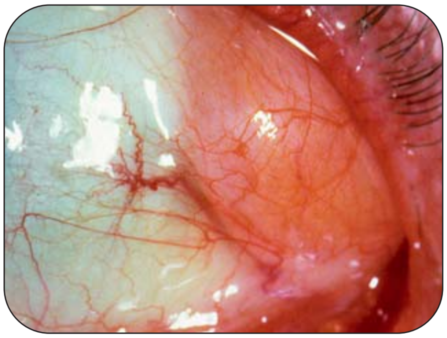

Fig. 3.87

Non-Hodgkin lymphoma and reactive lymphoid hyperplasia

Signs

- Smooth, fleshy, subconjunctival epibulbar (**Fig. 3.88**) or forniceal (**Fig. 3.89**) infiltrate.

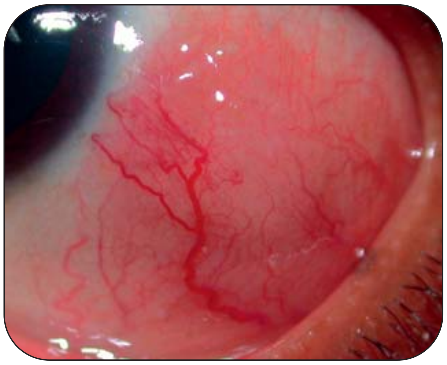

Fig. 3.88

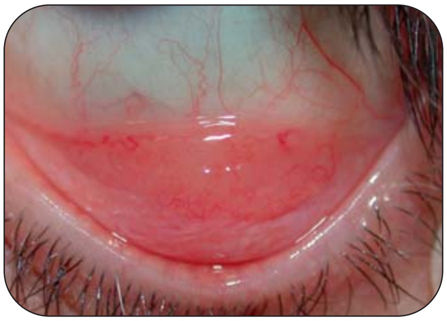

Fig. 3.89

Amyloidosis

Signs

- Firm yellowish subconjunctival deposits (**Fig. 3.90**).

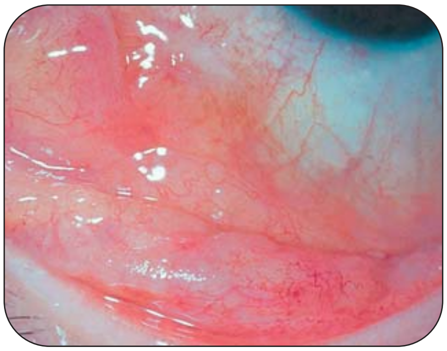

Fig. 3.90

Reactive pseudo-epitheliomatous hyperplasia

Cause

- Irritation or inflammation.

Signs

- Rapidly developing white hyperkeratotic mass with indistinct margins (**Fig. 3.91**).

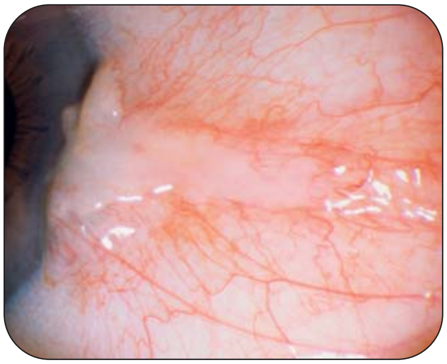

Fig. 3.91

Benign hereditary intraepithelial dyskeratosis

Signs

- Hyperplastic translucent lesion at the limbus associated with dilated vessels (**Fig. 3.92**).

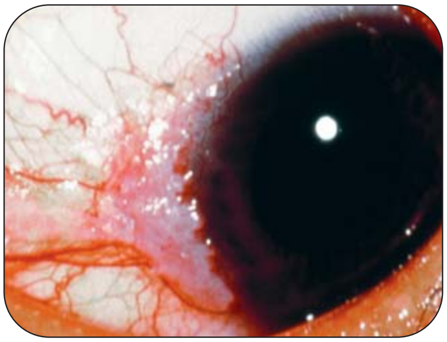

Fig. 3.92

Cystic

Primary retention cyst

Signs

- Thin-walled lesion containing clear fluid (**Fig. 3.93**).

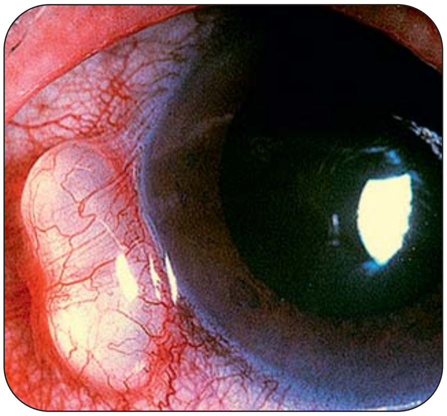

Fig. 3.93

Secondary inclusion cyst

Cause

- Strabismus surgery if the conjunctiva is not carefully sutured.
- Removal of pterygium.

Signs

- Smooth lesion that may be translucent (**Fig. 3.94**) or may contain turbid fluid (**Fig. 3.95**).

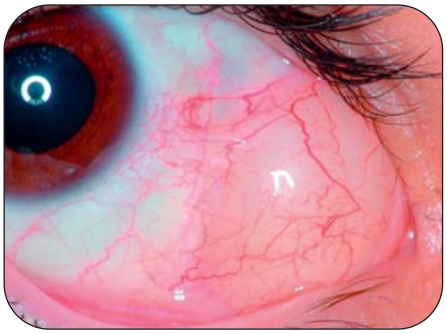

Fig. 3.94

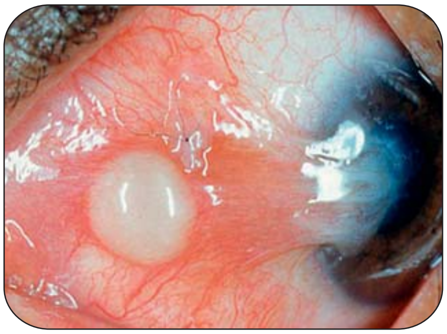

Fig. 3.95

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Episclera and sclera

4

ANTERIOR NON-NECROTISING DISEASE 93

Without a nodule 93

- *Diffuse (simple) episcleritis* 93
- *Diffuse scleritis* 93

With a nodule 94

- *Nodular episcleritis* 94
- *Nodular scleritis* 94

ANTERIOR NECROTISING DISEASE 95

- *Necrotising scleritis with inflammation* 95
- *Scleromalacia perforans* 95

POSTERIOR SCLERITIS 95

CAUSES AND SYSTEMIC ASSOCIATIONS OF SCLERITIS 96

- *Surgically-induced* 96
- *Herpes zoster ophthalmicus* 96
- *Rheumatoid arthritis* 96
- *Relapsing polychondritis* 96
- *Wegener granulomatosis* 97
- *Polyarteritis nodosa* 97
- *Systemic lupus erythematosus* 97

SCLERAL DISCOLORATION 98

Focal 98

- *Scleral hyaline plaque* 98
- *Alkaptonuria* 98
- *Systemic minocycline* 99

Diffuse blue 99

- *Osteogenesis imperfecta type 1 and 2* 99
- *Ehlers–Danlos syndrome type 6* 99
- *Pseudoxanthoma elasticum – AD type 2* 100
- *Turner syndrome* 100
- *Other uncommon syndromic associations* 100

ANTERIOR NON-NECROTISING DISEASE

Without a nodule

Diffuse (simple) episcleritis

Signs

- Interpalpebral hyperaemia involving superficial episcleral vessels (large tortuous and radially orientated) and deep episcleral vessels (smaller and interlacing).
- Normal vascular architecture even after multiple attacks.
- Tenon capsule is separated from the sclera by oedema.
- Involvement may be generalised (**Fig. 4.1**) or sectoral (**Fig. 4.2**).

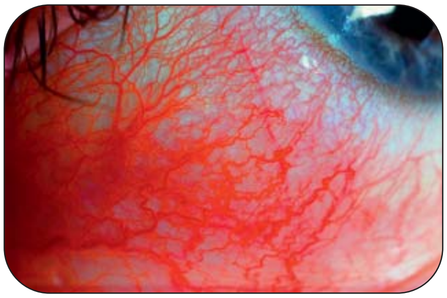

Fig. 4.1

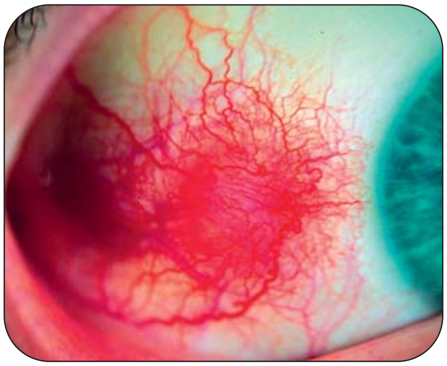

Fig. 4.2

Diffuse scleritis

Signs

- Swollen eyelids and tender globe.
- Intense hyperaemia and distortion of the deep episcleral vessels (**Fig. 4.3**).
- Variable involvement of the superficial episcleral and conjunctival vessels.
- The inflamed sclera has a violaceous hue when viewed in daylight.
- Involvement may be sectoral or generalised.

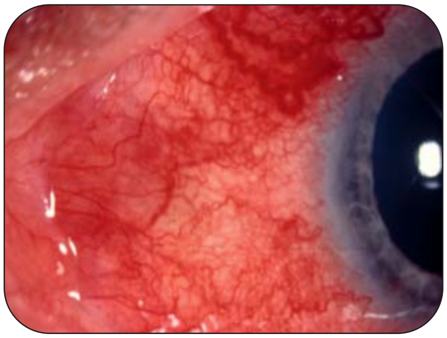

Fig. 4.3

With a nodule

Nodular episcleritis

Signs

- Tender interpalpebral nodule (**Fig. 4.4**) over which the conjunctiva can be moved, and the nodule itself can be moved slightly over the sclera.
- Dilatation of the conjunctival and superficial episcleral vessels which will blanch on instillation of 10% phenylephrine.
- A thin slit-lamp section shows that the deep beam is not displaced indicating that the scleral surface is flat (**Fig. 4.5**).

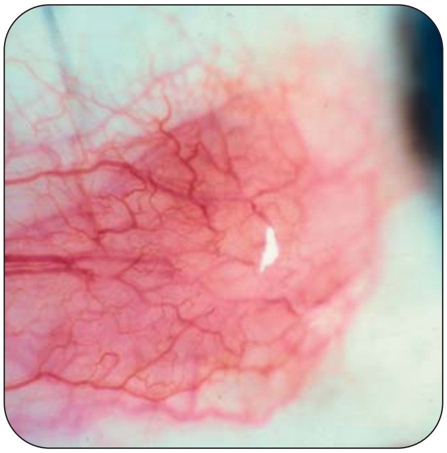

Fig. 4.4

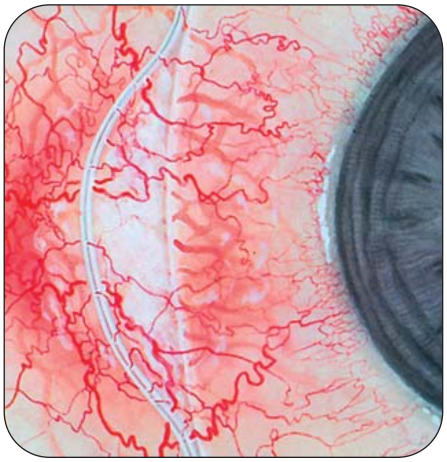

Fig. 4.5

Nodular scleritis

Signs

- Tender nodule which cannot be moved over the sclera (**Fig. 4.6A**).
- Dilatation of the conjunctival vessels, and the superficial and deep episcleral vasculature.
- Instillation of 10% phenylephrine constricts the conjunctival and superficial episcleral vessels but not the deep vessels (**Fig. 4.6B**).
- A thin slit-lamp section shows that the nodule displaces the entire beam because the scleral surface is elevated (**Fig. 4.7**).

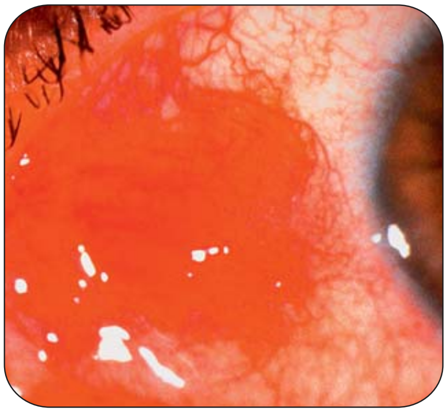

Fig. 4.6A

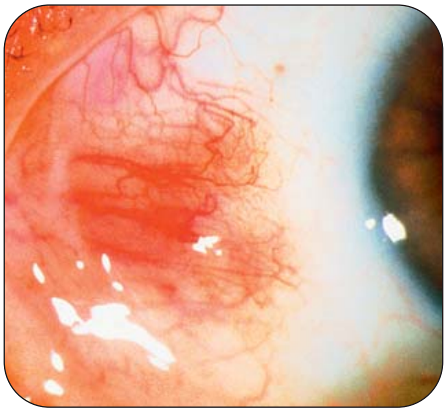

Fig. 4.6B

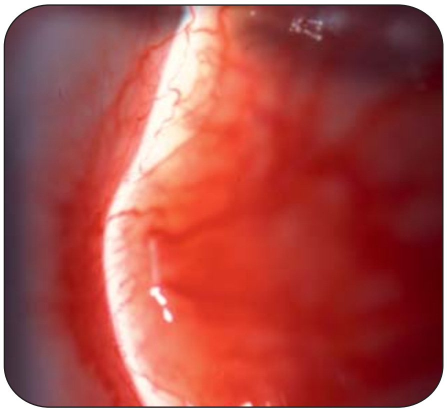

Fig. 4.7

ANTERIOR NECROTISING DISEASE

Necrotising scleritis with inflammation

Signs

- Vascular distortion and occlusion of the deep episcleral vessels.
- Painful avascular patches and necrotic areas (**Fig. 4.8**).
- Exposure of underlying uvea through thinned and necrotic sclera (**Fig. 4.9**).

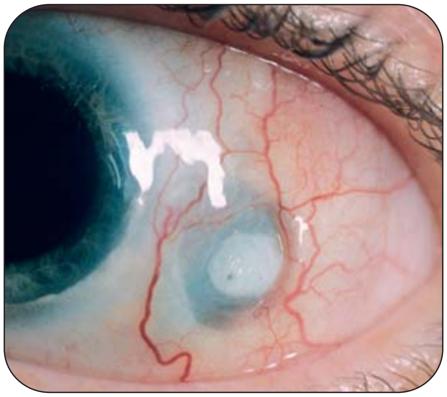
Fig. 4.8

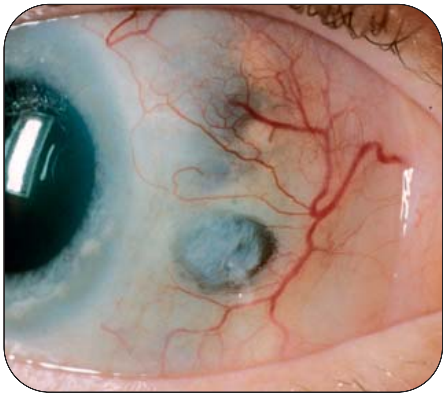
Fig. 4.9

Scleromalacia perforans

Definition

- An asymptomatic condition that is seen exclusively in patients with long-standing rheumatoid arthritis.

Signs

- Diffuse progressive scleral necrosis without inflammation resulting in exposure of large areas of underlying uvea (**Fig. 4.10**).

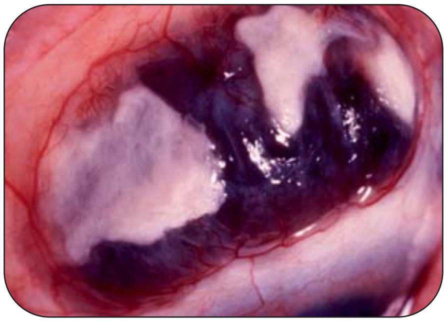
Fig. 4.10

POSTERIOR SCLERITIS

Signs

- Associated anterior scleritis in about 80% of cases.
- Orbital inflammation and proptosis.
- Defective ocular motility.
- Vitritis.
- Disc oedema.
- Choroidal ring detachment and chorioretinal folds.
- Macular oedema, exudative detachment, intraretinal deposits, subretinal exudation and a mass.
- Ultrasonography shows scleral thickening and fluid in sub-Tenon space giving rise to the characteristic ‘T’ sign in which the stem is formed by the optic nerve and the cross bar by the gap containing fluid (**Fig. 4.11**).
- CT shows scleral thickening and proptosis (**Fig. 4.12**).

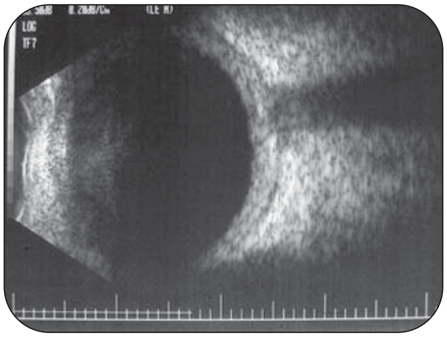
Fig. 4.11

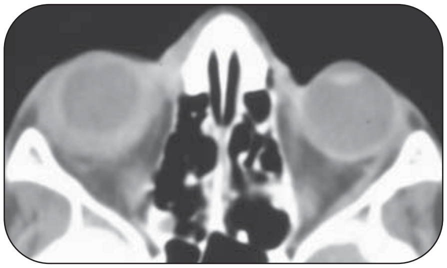
Fig. 4.12

CAUSES AND SYSTEMIC ASSOCIATIONS OF SCLERITIS

Surgically-induced

Presentation

Three weeks after a surgical procedure such as:

- Strabismus repair.
- Scleral buckling.
- Trabeculectomy (**Fig. 4.13**).
- Application of mitomycin C for the treatment of pterygium (**Fig. 4.14**).

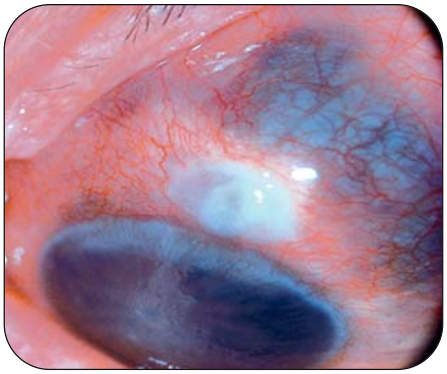

Fig. 4.13

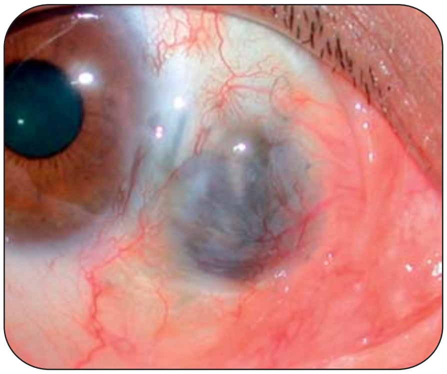

Fig. 4.14

Herpes zoster ophthalmicus

Signs

- Focal area of scleral atrophy (**Fig. 4.15**).

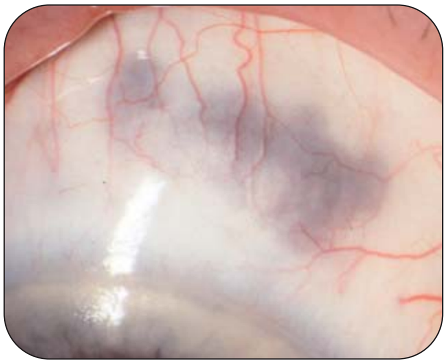

Fig. 4.15

Rheumatoid arthritis

Rheumatoid arthritis is by far the most common systemic association that may be associated with necrotising scleritis with inflammation (see below) as well as scleromalacia perforans (see Fig. 4.10).

Look for

- Symmetric, erosive arthritis involving the hands, ankles, knees and spine.
- Subcutaneous nodules (**Fig. 4.16**).
- Cutaneous vasculitis.

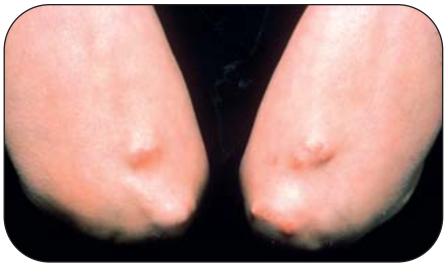

Fig. 4.16

Relapsing polychondritis

Polychondritis is often associated with intractable scleritis that may be non-necrotising or necrotising.

Look for

- Inflammation and destruction of the pinnae (**Fig. 4.17**) and a saddle-shaped nasal deformity (see Fig. 15.25).
- Stridor due to involvement of the larynx and trachea.
- Polyarthritis.

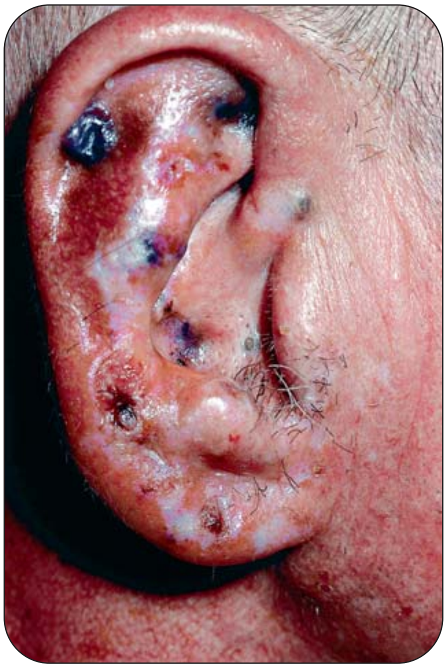

Fig. 4.17

Wegener granulomatosis

Wegener may be associated with rapidly progressive necrotising scleritis.

Look for

- Sinus inflammation and saddle-shaped nose.
- Lung cavitation (**Fig. 4.18**).
- Necrotising glomerulonephritis.

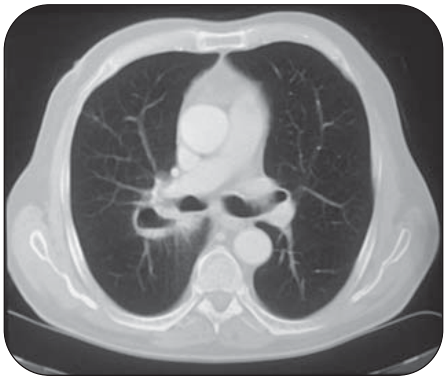

Fig. 4.18

Polyarteritis nodosa

Polyarteritis nodosa may be associated with aggressive necrotising disease which may precede systemic manifestations.

Look for

- Arthralgia and myalgia.
- Purpura (**Fig. 4.19**), easy bruising and cutaneous vasculitis.
- Renal and cardiovascular disease.

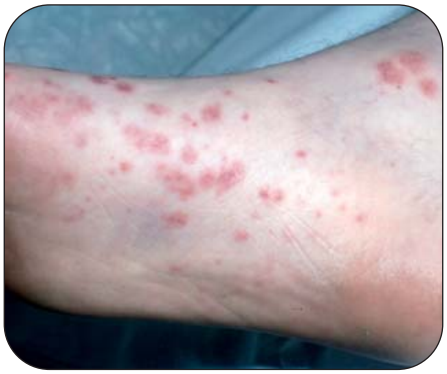

Fig. 4.19

Systemic lupus erythematosus

Lupus is occasionally associated with non-necrotising disease and rarely with necrotising scleritis.

Look for

- Facial rash with a characteristic 'butterfly' distribution (see Fig. 15.4).
- Cutaneous vasculitis and photosensitivity.
- Arthritis (**Fig. 4.20**).
- Glomerulonephritis.
- Hirsutism in females.

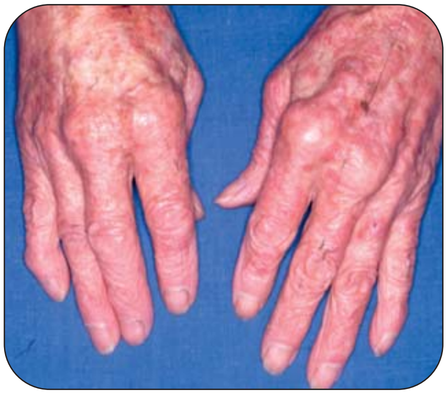

Fig. 4.20

SCLERAL DISCOLORATION

Focal

Scleral hyaline plaque

Definition

- A common, bilateral, innocuous, asymptomatic age-related condition.

Signs

- Dark-greyish, oval area close to the insertion of a horizontal rectus muscle (**Fig. 4.21**).

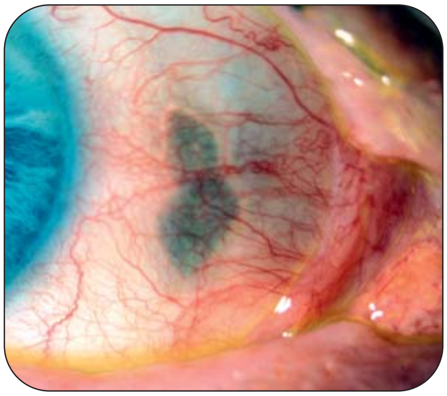

Fig. 4.21

Alkaptonuria

Definition

- A metabolic disorder that results in accumulation of black homogentistic acid in collagenous tissues (ochronosis).

Signs

- Interpalpebral, bluish-grey or black pigmentation of the conjunctiva, episclera, sclera and tendons of horizontal recti (**Fig. 4.22**).

Differential diagnosis

- Early scleromalacia perforans.

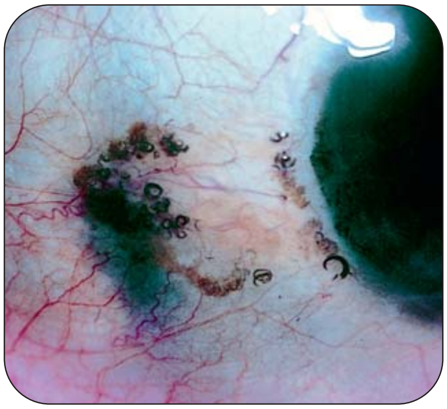

Fig. 4.22

Systemic minocycline

Signs

- Blue-grey paralimbal discoloration and involvement of the skin (see Fig. 1.53) may rarely occur.

Diffuse blue

Blue sclera is caused by thinning and transparency of scleral collagen fibres allowing visualisation of the underlying uvea (**Fig. 4.23**). It may be associated with the following systemic conditions.

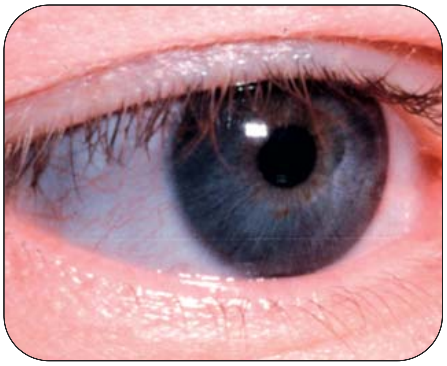

Fig. 4.23

Osteogenesis imperfecta type 1 and 2

Look for

- In type 1 – joint hypermobility and cardiac valve lesions.
- In type 2 – multiple fractures, short limbs (**Fig. 4.24**) and early demise.

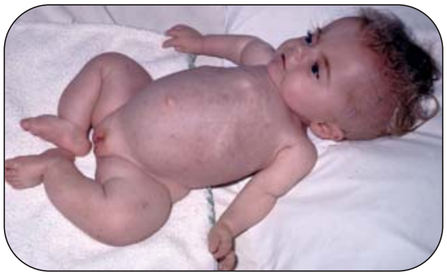

Fig. 4.24

Ehlers–Danlos syndrome type 6

Look for

- Thin and hyperelastic skin which bruises easily and heals slowly.
- Hyperextensible joints (**Fig. 4.25**) that easily dislocate.
- Dissecting aortic aneurysm and mitral valve disease.

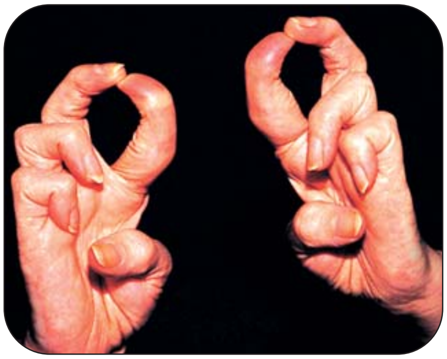

Fig. 4.25

Pseudoxanthoma elasticum – AD type 2

Look for

- Chicken skin cutaneous papules (**Fig. 4.26**) and hyperelasticity.
- High-arched palate.
- Mild angioid streaks.

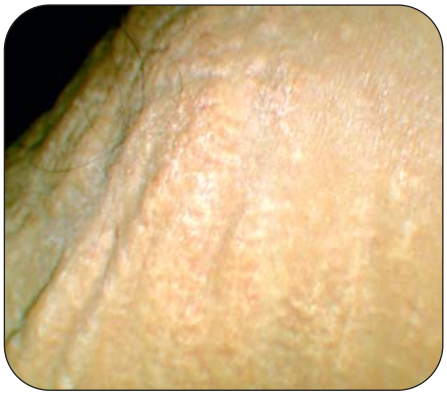

Fig. 4.26

Turner syndrome

Look for

- Sexual infantilism.
- Webbed neck (**Fig. 4.27**).
- Short stature with a broad chest.

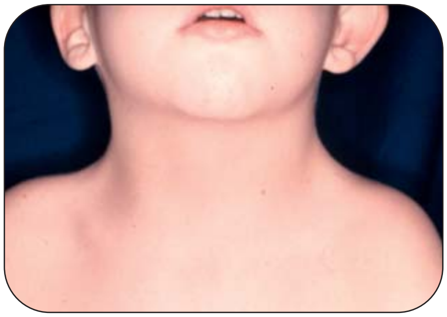

Fig. 4.27

Other uncommon syndromic associations

a. *Marshall–Smith.*

b. *Russell–Silver.*

c. *Roberts.*

d. *Marfan.*

e. *Hallermann–Streiff.*

f. *Bloch–Sulzberger* (incontinentia pigmenti).

5

Cornea

ABNORMAL SIZE **103**
Small **103**
- *Microcornea* *103*
- *Nanophthalmos* *103*
- *Microphthalmos* *103*

Large **104**
- *Megalocornea* *104*
- *Buphthalmos* *105*
- *High myopia* *105*

ABNORMAL SHAPE **106**
- *Keratoconus* *106*
- *Keratoglobus* *107*
- *Pellucid marginal degeneration* *107*
- *Posterior keratoconus* *107*
- *Cornea plana* *108*

PERINATAL CLOUDING **109**
- *Congenital glaucoma* *109*
- *Birth trauma* *109*
- *Intrauterine rubella* *109*
- *Sclerocornea* *109*
- *Peters anomaly* *110*
- *Congenital anterior staphyloma* *110*
- *Mucopolysaccharidoses* *110*
- *Mucolipidoses types II and IV* *111*
- *Congenital hereditary endothelial dystrophy* *111*
- *Congenital hereditary stromal dystrophy* *111*

PROMINENT NERVES **111**

EPITHELIAL MICROCYSTS AND VESICLES **112**
- *Epithelial oedema* *112*
- *Meesmann dystrophy* *112*
- *Cogan microcystic dystrophy* *112*

OEDEMA **113**
- *Iatrogenic endothelial damage* *113*
- *Hydrops* *113*
- *Fuchs endothelial dystrophy* *113*
- *Acute angle-closure* *114*
- *Disciform keratitis* *114*
- *Acute postoperative endophthalmitis* *114*
- *Congenital hereditary endothelial dystrophy* *114*

PUNCTATE LESIONS **115**
- *Punctate epithelial erosions* *115*
- *Superficial punctate keratitis* *115*
- *Punctate epithelial keratitis* *115*
- *Nummular keratitis* *116*

FILAMENTS **116**
- *Keratoconjunctivitis sicca* *116*
- *Superior limbic keratoconjunctivitis* *117*

EPITHELIAL DEFECTS AND ULCERS **117**
Dendritic and pseudodendritic ulceration **117**
- *Herpes simplex dendritic ulcer* *117*
- *Herpes zoster pseudodendrite* *118*
- *Acanthamoeba pseudodendrite* *118*
- *Other causes of pseudodendrites* *118*

Large epithelial defects **119**
- *Corneal abrasion* *119*
- *Herpes simplex geographic ulcer* *119*
- *Neurotrophic keratopathy* *119*
- *Exposure keratopathy* *120*
- *Epithelial macroerosion* *120*

Microbial ulcers **121**
- *Bacterial keratitis* *121*
- *Candida keratitis* *121*
- *Filamentous keratitis* *121*
- *Acanthamoeba keratitis* *122*

EPITHELIAL DEPOSITS **123**
- *Vortex keratopathy* *123*
- *Adrenochrome* *123*
- *Fluoroquinolones* *123*
- *Epithelial iron lines* *123*

DEGENERATIONS IN BOWMAN LAYER AND ANTERIOR STROMA **124**
- *Band keratopathy* *124*
- *Spheroidal degeneration* *124*
- *Lipid keratopathy* *125*
- *Salzmann nodular degeneration* *125*
- *Infectious crystalline keratopathy* *125*

DYSTROPHIES OF BOWMAN LAYER AND ANTERIOR STROMA **126**
- *Reis–Bückler dystrophy (Bowman layer 1)* *126*
- *Thiel–Behnke dystrophy (Bowman layer 2)* *126*
- *Central Schnyder (crystalline) dystrophy* *126*
- *Gelatinous drop-like dystrophy* *127*

DEPOSITS IN STROMA AND DESCEMET MEMBRANE **127**
Metabolic **127**
- *Cystinosis* *127*
- *Kayser–Fleischer ring* *127*
- *Immunoprotein deposits* *128*
- *Mucopolysaccharidoses* *128*
- *Hypoproteinaemias* *128*

Non-metabolic **129**
- *Chrysiasis* *129*
- *Argyrosis* *129*
- *Phenothiazines* *129*
- *Bietti crystalline dystrophy* *130*
- *Cornea farinata* *130*

STROMAL DYSTROPHIES **130**
- *Lattice dystrophy type 1 (Biber–Haab–Dimmer)* *130*
- *Lattice dystrophy type 2 (Meretoja syndrome)* *130*
- *Lattice dystrophy type 3* *131*
- *Granular dystrophy type 1* *131*
- *Granular dystrophy type 2 (Avellino)* *131*
- *Macular dystrophy* *132*
- *Central cloudy dystrophy of François* *132*
- *Fleck dystrophy* *132*

INTERSTITIAL KERATITIS **133**
Signs **133**
Systemic associations **133**
- *Congenital syphilis* *133*
- *Acquired syphilis* *133*
- *Lyme disease (stage 3)* *134*
- *Lymphogranuloma venereum* *134*
- *Cogan syndrome* *134*
- *Miscellaneous* *134*

PERIPHERAL LESIONS 134

Circumcorneal opacification 134
- *Vogt white limbal girdle 134*
- *Corneal arcus 135*
- *Pseudogerontoxon (anterior embryotoxon) 135*
- *Posterior embryotoxon 135*
- *Kayser–Fleischer ring 135*

Infiltrates 136
- *Marginal keratitis 136*
- *Rosacea keratitis 136*
- *Chlamydial keratitis 136*
- *Phlyctenulosis 137*
- *Reiter syndrome 137*
- *Exposure keratopathy 137*
- *Sclerosing keratitis 137*
- *Acute stromal keratitis 138*
- *Inflammatory bowel disease 138*

Thinning and ulceration 138
- *Furrow degeneration 138*
- *Dellen 138*
- *Terrien marginal degeneration 139*
- *Mooren ulcer 139*
- *Rheumatoid arthritis 140*
- *Other diseases 140*

ABNORMAL SIZE

Small

Microcornea

Definition

- A very rare, congenital, unilateral or bilateral AD or AR condition that has many ocular and systemic syndromic associations (Table 5.1).

Signs

- Adult horizontal corneal diameter is 10 mm or less (**Fig. 5.1**).
- Shallow anterior chamber.
- Other ocular dimensions are normal.

Table 5.1 **Associations of microcornea**

1. **Ocular**
    - Cornea plana
    - Corneal leukoma
    - Iris abnormalities
    - Microphakia
    - Congenital cataract
    - Glaucoma
2. **Syndromic**
    - Turner
    - Ehlers–Danlos
    - Weill–Marchesani
    - Waardenburg
    - Nance–Horan
    - Cornelia de Lange

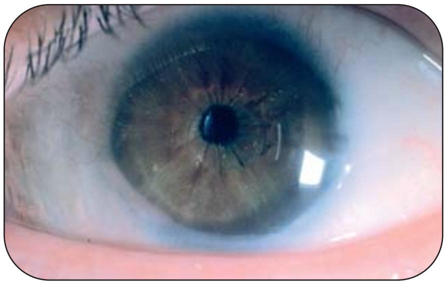

Fig. 5.1

Nanophthalmos

Definition

- A rare, bilateral, congenital condition in which the globes have reduced volume but are otherwise grossly normal.

Signs

- Very high hypermetropia (**Fig. 5.2**).
- Adult corneal diameter is reduced but the lens has a normal volume.
- Short axial length averaging 18 mm.
- Shallow anterior chamber.
- Thick sclera.
- Fundus may show a crowded disc, vascular tortuosity and macular hypoplasia.

Late problems

- Angle-closure glaucoma.
- Uveal effusion characterised by choroidal detachment and retinal detachment (**Fig. 5.3**).
- Retinal detachment.
- Poorly tolerated intraocular surgery.

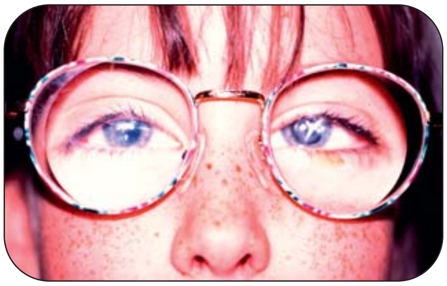

Fig. 5.2

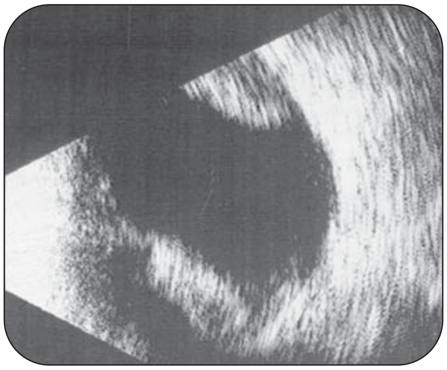

Fig. 5.3

Microphthalmos

Definition

- An uncommon, unilateral or bilateral condition in which the axial length of the globe is reduced.

Types

a. *Non-colobomatous* (simple – **Fig. 5.4** and Table 5.2).

b. *Colobomatous* (**Fig. 5.5** and Table 5.3).

Table 5.2 **Classification of non-colobomatous microphthalmos**

1. **Isolated**
    - Sporadic
    - Inherited (AD, AR, XL-R)
    - With persistent anterior fetal vasculature (see Fig. 9.19)
2. **Microphthalmos with cyst** (see Fig. 2.28)
3. **Intrauterine infections**
    - Rubella
    - Toxoplasmosis
    - Cytomegalovirus
    - Varicella

Table 5.3 **Classification of colobomatous microphthalmos**

1. **Isolated**
    - Sporadic
    - Inherited (AD)
2. **Syndromic associations**
    - Patau (trisomy 13)
    - Edward (trisomy 18)
    - Cat-eye (partial trisomy 22)
    - Pagon (CHARGE)
    - Meckel-Gruber
    - Lenz microphthalmos
    - Temple-Al Gazali
    - Delleman
    - Gorlin-Goltz

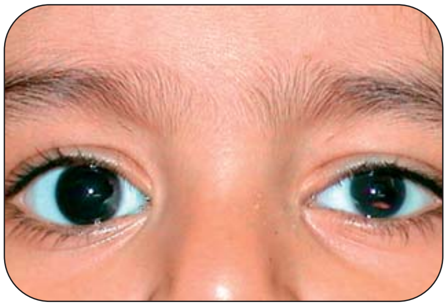

Fig. 5.4

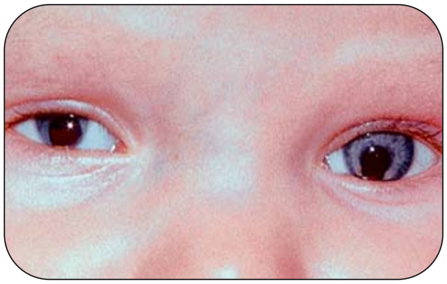

Fig. 5.5

Large

Megalocornea

Definition

- A rare, congenital, bilateral, non-progressive usually XL-R condition that has many systemic associations (Table 5.4).

Table 5.4 **Systemic associations of megalocornea**

- Marfan syndrome
- Apert syndrome
- Ehlers-Danlos syndrome
- Down syndrome
- Osteogenesis imperfecta
- Progressive facial hemiatrophy
- Renal carcinoma
- Mental retardation

Signs

- Clear cornea with a horizontal diameter of 13 mm or more (**Fig. 5.6**).
- Very deep anterior chamber.
- Normal intraocular pressure.
- Pigment dispersion.
- High myopia and astigmatism but good visual acuity.
- Lens subluxation may occur as a result of zonular stretching.

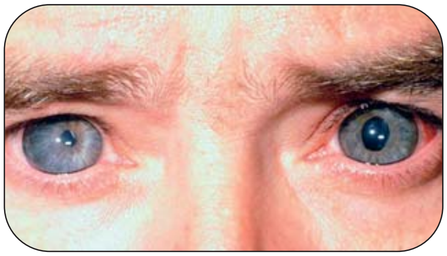

Fig. 5.6

Buphthalmos

Definition

An uncommon, usually bilateral, condition in which the globe is large due to increased intraocular pressure during the first 3 years of life. It has many ocular and systemic associations (Table 5.5).

Signs

- Large cornea with variable oedema and scarring (**Fig. 5.7**).
- Very deep anterior chamber.
- Angle anomalies.
- Horizontal ruptures in Descemet membrane (Haab striae).
- Myopia.
- Optic disc cupping.

Table 5.5 **Associations of buphthalmos**

1. **Ocular**
    - Axenfeld–Rieger anomaly
    - Aniridia
    - Peters anomaly
    - Congenital ectropion uveae
2. **Systemic**
    - Lowe syndrome
    - Sturge–Weber syndrome
    - Down syndrome
    - NF1
    - Rieger syndrome
    - Rubinstein–Taybi syndrome
    - Pierre Robin syndrome
    - Patau syndrome (trisomy 13)
    - Naevus of Ota
    - Mucopolysaccharidoses
    - Marfan syndrome

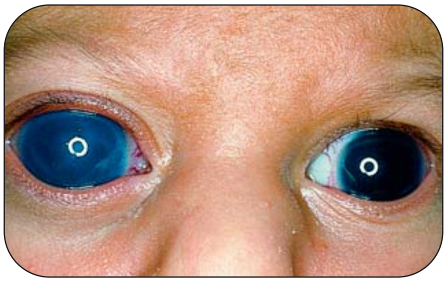

Fig. 5.7

High myopia

Definition

- Refraction is greater than –6D and axial length is greater than 25 mm.

Signs

- Enlargement of the globe and cornea (**Fig. 5.8**).
- Chorioretinal degeneration and maculopathy.
- Predisposition to retinal detachment and glaucoma.
- Staphyloma.

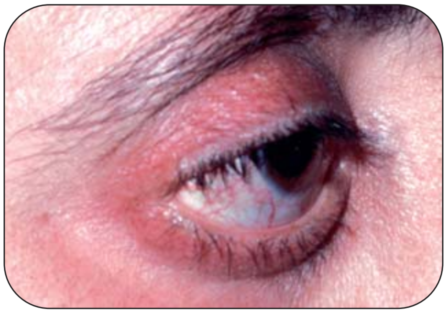

Fig. 5.8

Look for

- Stickler syndrome.
- Cohen syndrome.
- Marfan syndrome.
- Ehlers–Danlos syndrome.
- Fabry disease.

ABNORMAL SHAPE

Keratoconus

Definition

- A common, acquired condition which is bilateral in 85% of cases, although the severity of involvement may be asymmetric.
- It is associated with many ocular and systemic conditions (Table 5.6).

Signs

- Central or paracentral stromal thinning with inferior apical protrusion (**Fig. 5.9**).
- Munson sign – bulging of the lower lid in downgaze (**Fig. 5.10A** right eye; left eye is normal – **Fig. 5.10B**).
- Vogt striae – fine deep vertical stromal stress lines which temporarily disappear on digital pressure (**Fig. 5.11**).
- Acute hydrops – corneal oedema resulting from tears in Descemet membrane (**Fig. 5.12**).
- Variable corneal scarring, depending on severity (**Fig. 5.13**).

Table 5.6 **Associations of keratoconus**

1. **Ocular**
   - Vernal disease
   - Blue sclera
   - Prominent corneal nerves
   - Aniridia
   - Ectopia lentis
   - Retinitis pigmentosa
   - Leber congenital amaurosis
2. **Systemic**
   - Down syndrome
   - Ehlers–Danlos syndrome type 6
   - Marfan syndrome
   - Apert syndrome
   - Turner syndrome
   - Atopic dermatitis
   - Osteogenesis imperfecta type 1
   - Mitral valve prolapse

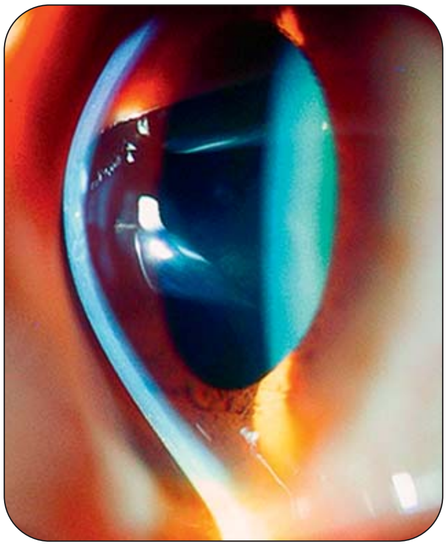
Fig. 5.9

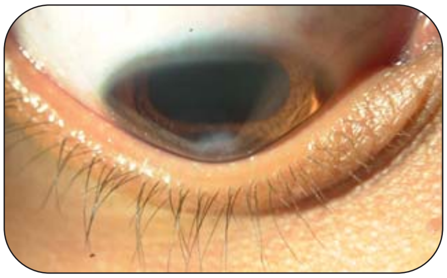
Fig. 5.10A

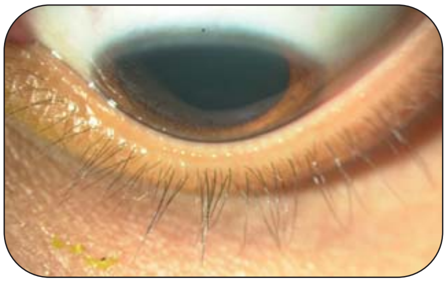
Fig. 5.10B

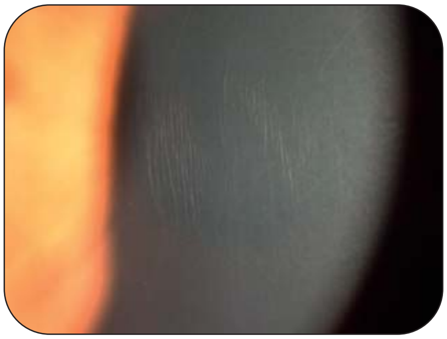
Fig. 5.11

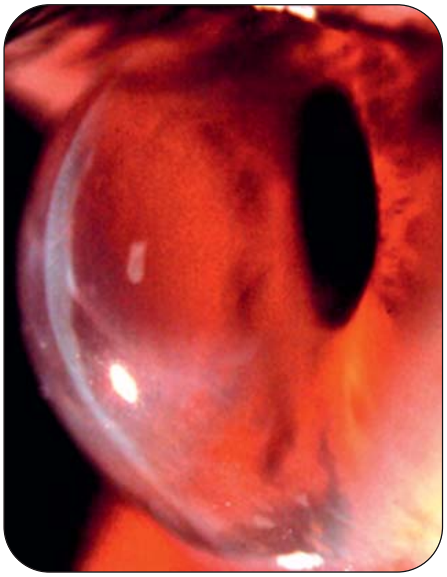
Fig. 5.12

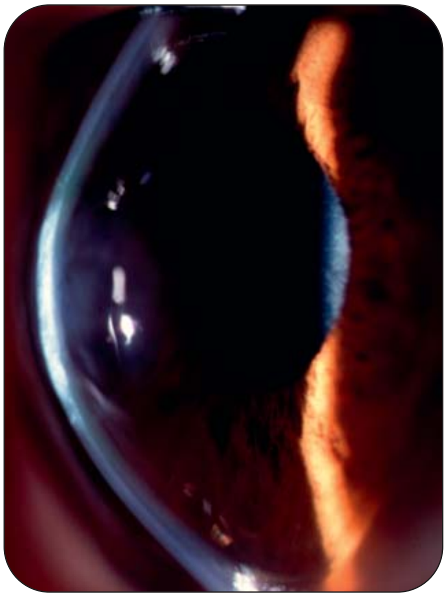
Fig. 5.13

Keratoglobus

Definition

- A very rare, bilateral, congenital condition.

Signs

- Mid-peripheral thinning, resulting in protrusion of the entire cornea (**Fig. 5.14**).
- Very deep anterior chamber.
- Acute hydrops in advanced cases.

Look for

- A syndrome comprising blue sclera, hyperextensible joints and dental abnormalities.

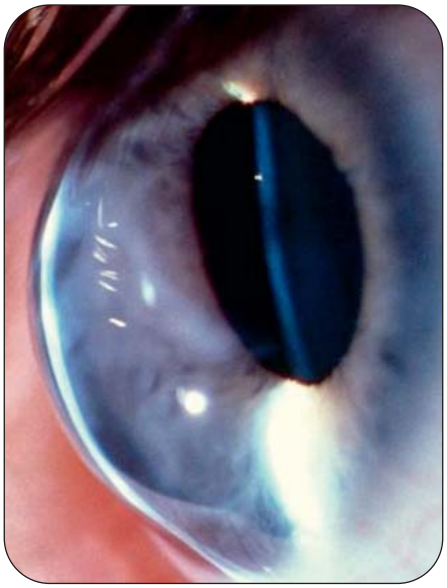

Fig. 5.14

Pellucid marginal degeneration

Definition

- An uncommon, acquired, bilateral condition.

Signs

- High astigmatism.
- Inferior, crescent-shaped area of peripheral corneal thinning, 1–2 mm in width, extending from the 4 o'clock to 8 o'clock positions, which is separated from the limbus by normal cornea (**Fig. 5.15**).
- Protrusion located above the area of thinning.

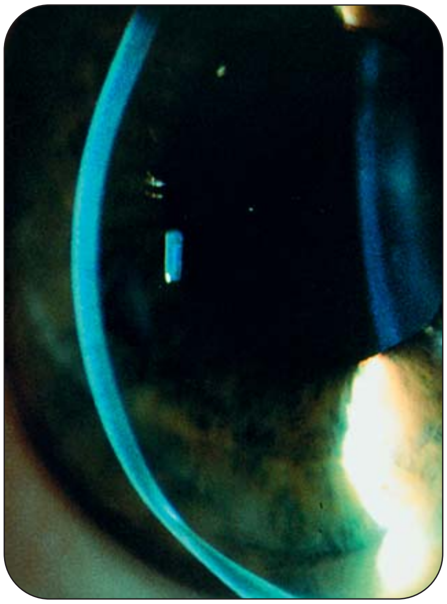

Fig. 5.15

Posterior keratoconus

Definition

- A very rare, usually unilateral developmental anomaly that bears no relationship to anterior keratoconus.

Signs

a. *Generalis* – increase in curvature of the entire posterior corneal surface.

b. *Conscriptus* – localised central posterior corneal indentation, often associated with opacification (**Fig. 5.16**).

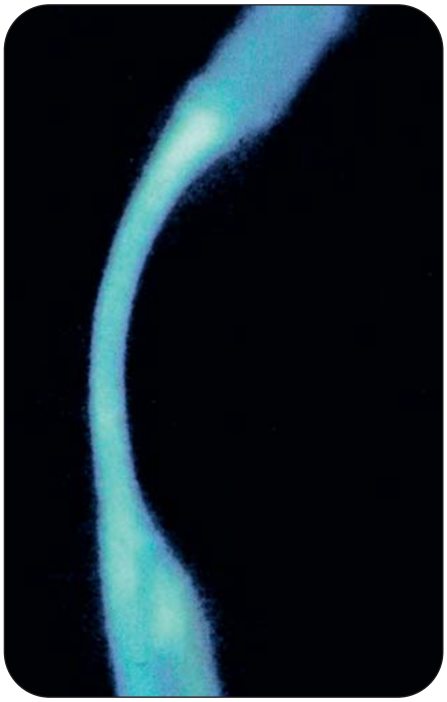

Fig. 5.16

Cornea plana

Definition

- A very rare bilateral, congenital condition which never occurs in isolation.

Signs

- Hypermetropia.
- Severe decrease in corneal curvature (**Fig. 5.17**).
- Shallow anterior chamber.

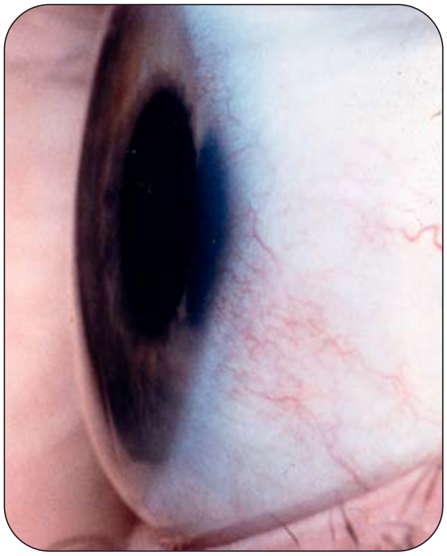

Fig. 5.17

PERINATAL CLOUDING

Congenital glaucoma

Signs

- Unilateral or bilateral corneal oedema (**Fig. 5.18**).

Look for

- Buphthalmos.
- Ruptures in Descemet membrane (Haab striae) that may be horizontal or concentric with the limbus.

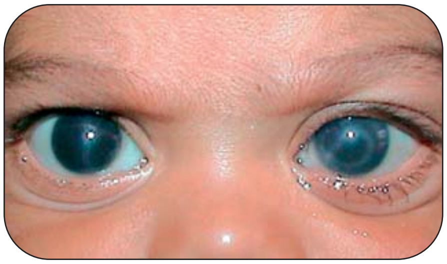

Fig. 5.18

Birth trauma

Signs

- Linear usually vertical or oblique ruptures in Descemet membrane associated with oedema (**Fig. 5.19**).

Look for

- Forceps marks on the eyelids or cheeks.

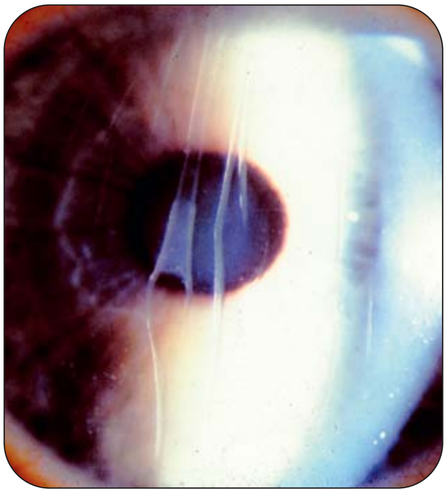

Fig. 5.19

Intrauterine rubella

Intrauterine rubella may cause corneal clouding as well as microphthalmos, cataract, anterior uveitis, retinitis and strabismus.

Sclerocornea

Definition

- A rare, congenital, usually bilateral but asymmetric condition.

Signs

- Opacification and vascularisation of the peripheral or entire cornea (**Fig. 5.20**).
- If only the peripheral cornea is involved, the resulting ‘scleralisation’ makes the cornea appear smaller than normal (**Fig. 5.21**).

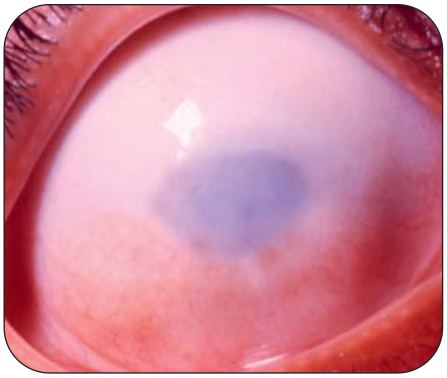

Fig. 5.20

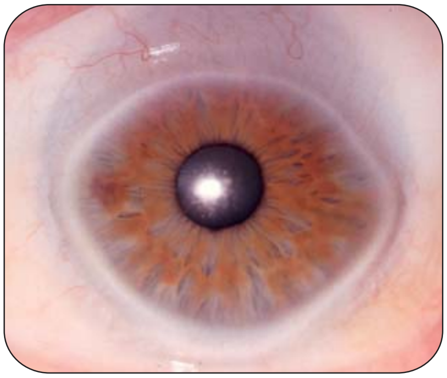

Fig. 5.21

Look for

- Peters anomaly (see next).
- Cornea plana.

Peters anomaly

Definition

- A usually bilateral, congenital condition that varies in severity.

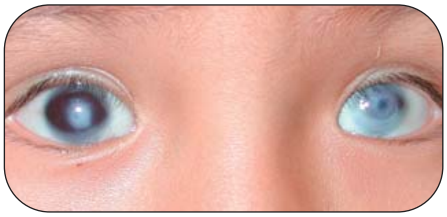

Fig. 5.22

Signs

- Central corneal opacities asymmetric in extent (**Fig. 5.22**) and an underlying defect of the posterior stroma, Descemet membrane and endothelium.
- May be associated with keratolenticular or iridocorneal adhesions best detected by ultrasonography (**Fig. 5.23**).

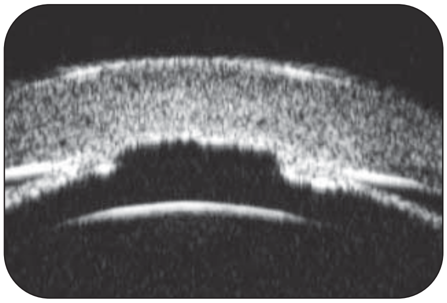

Fig. 5.23

Congenital anterior staphyloma

Definition

- A very rare, congenital, usually unilateral condition.

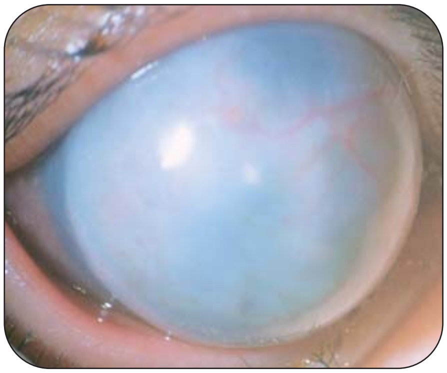

Fig. 5.24

Signs

- Opaque cornea lined posteriorly with uveal tissue that protrudes beyond the eyelids (**Fig. 5.24**).
- Keratectasia is identical except that it is not lined with uveal tissue.

Mucopolysaccharidoses

Mucopolysaccharidoses that occasionally may be associated with congenital corneal clouding include:

a. *Hurler syndrome (MPS I-H).*

b. *Scheie (MPS I-S) syndrome.*

c. *Morquio syndrome (MPS types IVA and IVB).*

d. *Maroteaux–Lamy syndrome (types VIA and VIB).*

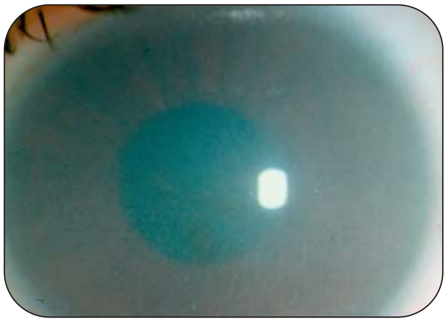

Fig. 5.25

Signs

- Diffuse corneal clouding caused by fine punctate grey opacities (**Fig. 5.25**).

Mucolipidoses types II and IV

These are frequently associated with congenital corneal opacification.

Congenital hereditary endothelial dystrophy

Presentation

- At or soon after birth with bilateral symmetric diffuse corneal oedema.

Signs

- The appearance may vary from a blue-grey ground-glass appearance (**Fig. 5.26**) to total opacification (**Fig. 5.27**).

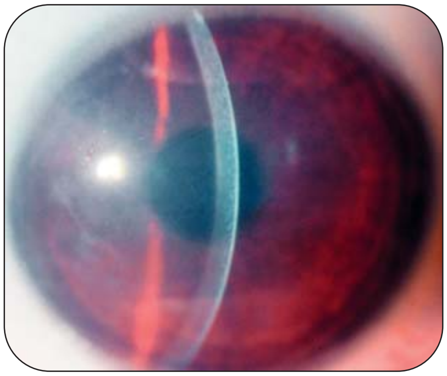

Fig. 5.26

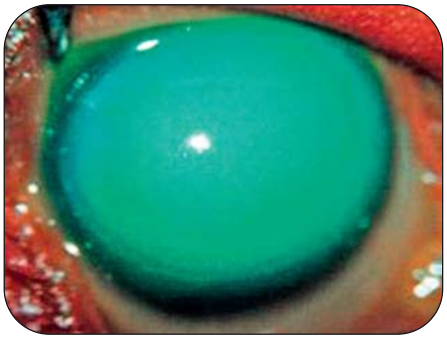

Fig. 5.27

Congenital hereditary stromal dystrophy

Definition

- A very rare, non-progressive AD condition.

Signs

- Diffuse central haze composed of flaky-feathery opacities primarily involving the central anterior stroma.

PROMINENT NERVES

Signs

- Linear branching lines within the corneal stroma (**Figs 5.28 and 5.29**).

Causes

- See Table 5.7.

Table 5.7 **Causes of prominent nerve fibres**

1. **Local**
    - Fuchs dystrophy
    - Keratoconus
    - Posterior polymorphous dystrophy
2. **Systemic**
    - Primary amyloidosis
    - Multiple endocrine neoplasia type II a and b
    - NF1
    - Refsum disease
    - Leprosy
    - Hereditary ichthyosis

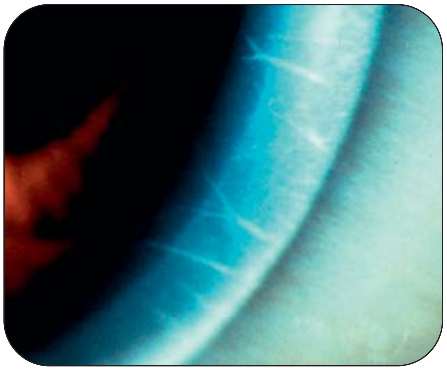

Fig. 5.28

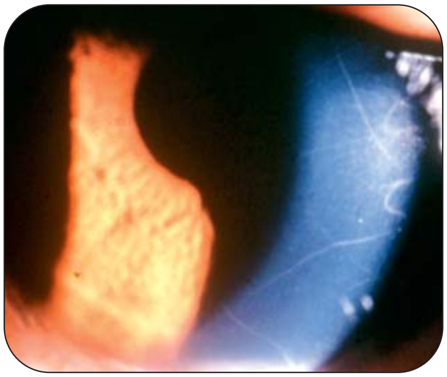

Fig. 5.29

EPITHELIAL MICROCYSTS AND VESICLES

Epithelial oedema

Signs

- Loss of normal corneal lustre (**Fig. 5.30**) which may be associated with vesicles and bullae (**Fig. 5.31**).

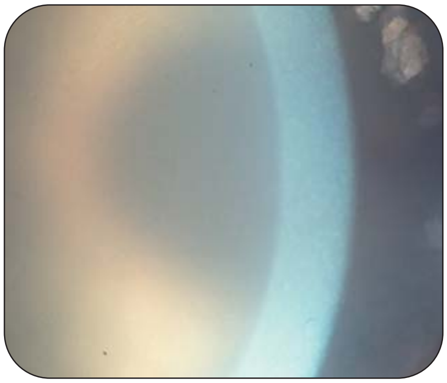

Fig. 5.30

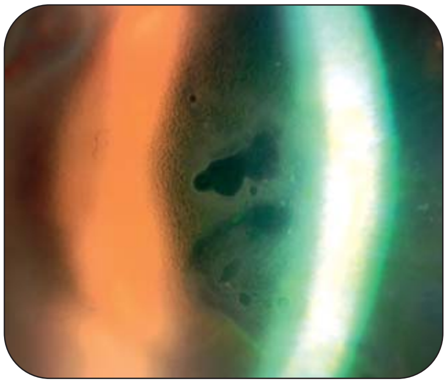

Fig. 5.31

Meesmann dystrophy

Definition

- A very rare, innocuous AD condition.

Signs

- Myriads of tiny epithelial cysts most numerous in the interpalpebral region.
- The lesions appear grey in direct illumination (**Fig. 5.32**) but clear on retroillumination.

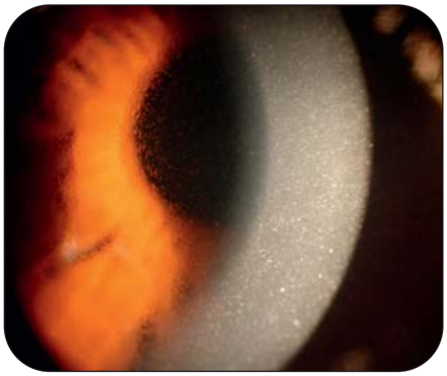

Fig. 5.32

Cogan microcystic dystrophy

Definition

- A common condition which is frequently associated with bilateral spontaneous recurrent corneal erosion.

Signs

- Sparse microcysts, dots and fingerprint-like, or map-like epithelial lesions seen singly or in various combinations (**Fig. 5.33**).

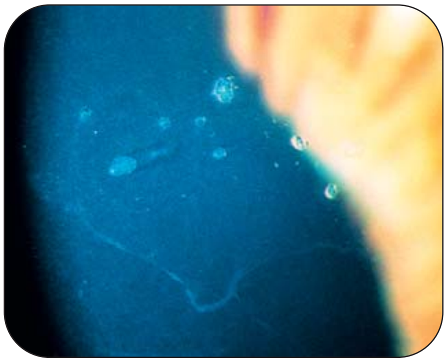

Fig. 5.33

OEDEMA

Corneal oedema is a common condition that may be caused by endothelial disease or severe elevation of intraocular pressure in an eye with a normal endothelium.

Iatrogenic endothelial damage

Cause

- Damage may occur during cataract surgery, particularly when associated with problems with lens implantation (pseudophakic bullous keratopathy – **Fig. 5.34**).

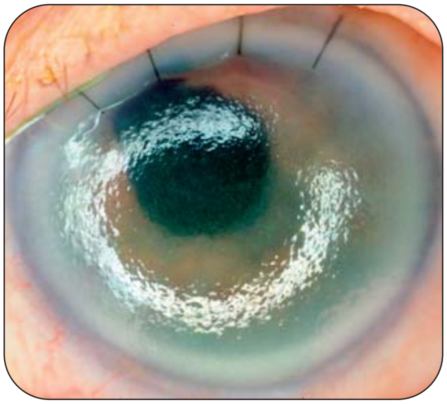

Fig. 5.34

Hydrops

Causes

- Advanced keratoconus (see Fig. 5.12) and occasionally keratoglobus (**Fig. 5.35**).

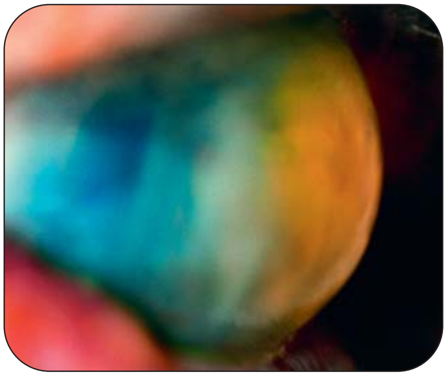

Fig. 5.35

Fuchs endothelial dystrophy

Definition

- An uncommon bilateral condition which usually occurs in old age.

Signs

- Starts as tiny central excrescences of Descemet membrane (cornea guttata – **Fig. 5.36**) which gradually become confluent.
- Subsequent endothelial dysfunction results in stromal oedema and bullous keratopathy.

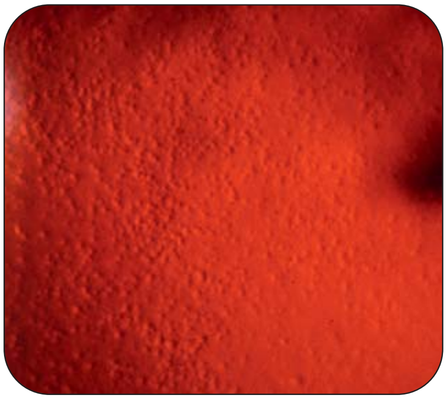

Fig. 5.36

Acute angle-closure

Signs

- Corneal oedema.
- Severe elevation of intraocular pressure.
- Ciliary injection.
- Fixed dilated pupil.
- Shallow anterior chamber (**Fig. 5.37**).
- Closed angle on gonioscopy.

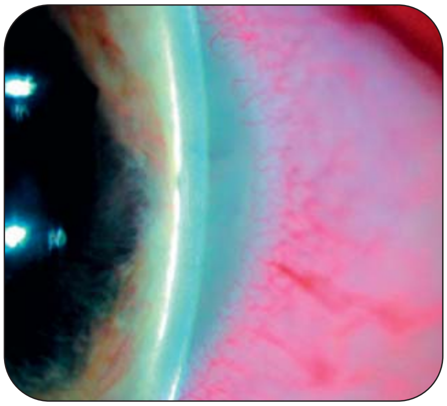

Fig. 5.37

Disciform keratitis

Definition

- A unilateral condition associated with herpes simplex and occasionally herpes zoster.

Signs

- Oval area of stromal and epithelial oedema with underlying keratic precipitates (**Fig. 5.38**).
- Surrounding ring of stromal opacity (Wessely ring) may be seen in long-standing cases.

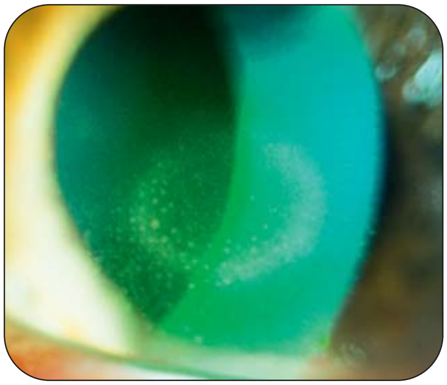

Fig. 5.38

Acute postoperative endophthalmitis

Signs

- Endophthalmitis is frequently associated with corneal oedema (**Fig. 5.39**).

Look for

- Conjunctival injection and discharge.
- Fibrinous anterior uveitis and enlarging hypopyon.
- Impaired red reflex.
- Vitreous cells and debris.

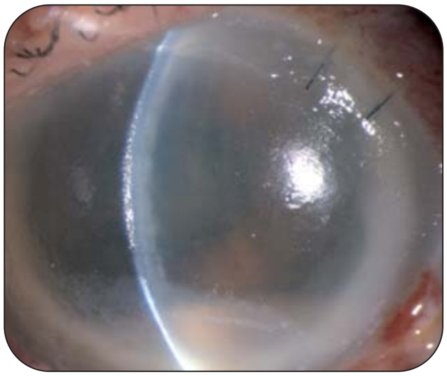

Fig. 5.39

Congenital hereditary endothelial dystrophy

- See Figs 5.36 and 5.37.

PUNCTATE LESIONS

Punctate epithelial erosions (PEE)

Definition

- A common, non-specific, unilateral or bilateral, condition with many diverse causes.

Signs

- Tiny non-specific epithelial defects that stain with fluorescein (**Fig. 5.40**) and rose Bengal.

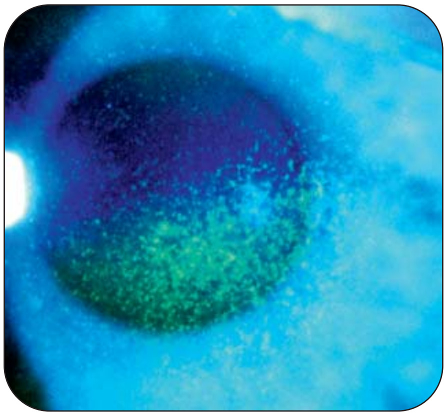

Fig. 5.40

Causes

Maximal intensity of may indicate the probably aetiology as follows:

a. *Superior* – vernal disease, superior limbic keratoconjunctivitis, floppy eyelids, poorly fitting contact lenses, chlamydial infection and subtarsal foreign body.

b. *Interpalpebral* – dry eye, reduced corneal sensation and exposure to ultraviolet light.

c. *Inferior* – chronic blepharitis, lagophthalmos, trichiasis, rosacea, toxicity from drops and self-induced.

Superficial punctate keratitis (SPK)

Definition

- SPK is the hallmark of Thygeson disease which is an idiopathic usually bilateral condition that typically affects young adults.

Signs

- Coarse, distinct, granular, greyish, elevated epithelial lesions that may be associated with a mild subepithelial haze (**Fig. 5.41**).

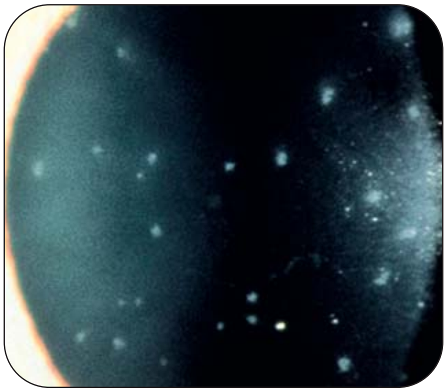

Fig. 5.41

Punctate epithelial keratitis (PEK)

Definition

- A unilateral or bilateral condition with many diverse causes.

Signs

- Small, multiple, frequently bilateral subepithelial opacities (**Fig. 5.42**) that stain poorly with fluorescein but well with rose Bengal.

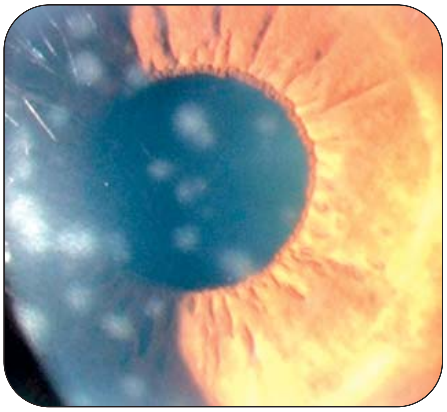

Fig. 5.42

Causes

- Adenovirus infection.
- Herpes zoster ophthalmicus.
- Reiter disease.

- Hypersensitivity to contact lens solution.
- Epithelial corneal graft rejection in donor cornea.
- Microsporidia infection.

Nummular keratitis

Definition

- An uncommon, unilateral or bilateral, condition caused by a variety of infections.

Signs

- Large, round, granular subepithelial deposits just beneath Bowman layer, surrounded by a halo of stromal haze (**Fig. 5.43**).

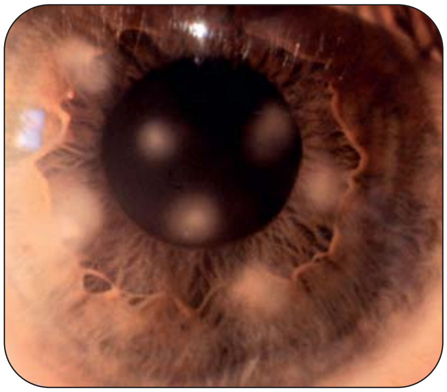

Fig. 5.43

Causes

- Herpes zoster ophthalmicus.
- Epstein–Barr virus infection.
- Acanthamoeba infection.
- Lyme disease.
- Onchocerciasis.
- Brucellosis.
- Dimmler nummular keratitis.

FILAMENTS

Keratoconjunctivitis sicca (KCS)

Definition

- A common, bilateral condition which may occur in isolation or in association with a systemic disorder (Tables 5.8 and 5.9).

Table 5.8 **Systemic associations of KCS**

1. **Autoimmune disorders**
    - Primary Sjögren syndrome
    - Rheumatoid arthritis
    - Systemic lupus erythematosus
    - Scleroderma
    - Polymyositis and dermatomyositis
    - Polyarteritis nodosa
    - Psoriatic arthritis
    - Chronic primary biliary cirrhosis
    - Hashimoto thyroiditis
    - Hypergammaglobulinaemia
    - Waldenström macroglobulinaemia
    - Lymphocytic interstitial pneumonitis
    - Interstitial nephritis
2. **Infiltrative disorders**
    - Sarcoidosis
    - Lymphoma
    - Haemochromatosis
    - Amyloidosis

Table 5.9 **Miscellaneous causes of filamentary keratitis**

1. **Ocular**
    - Recurrent erosions
    - Eye patching
    - Exposure keratopathy
    - Neurotrophic keratopathy
    - Herpes zoster ophthalmicus
2. **Systemic**
    - Essential blepharospasm
    - Midbrain strokes
3. **Drug-associated**
    - Clonidine
    - Propanolol
    - Amitriptyline

Signs

- Small, mucous filaments attached to the corneal surface (**Fig. 5.44**).
- Staining of the filaments and interpalpebral conjunctiva with rose Bengal (**Fig. 5.45**).
- Corneal melting in severe cases (**Fig. 5.46**).

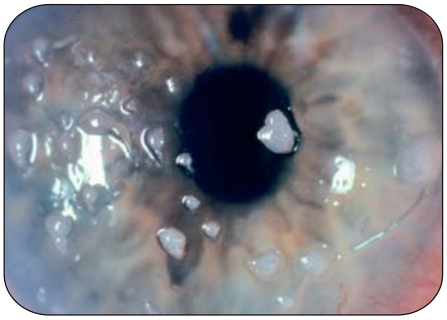

Fig. 5.44

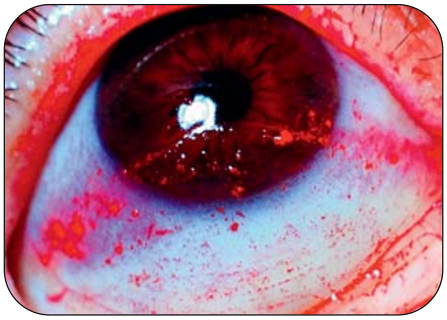

Fig. 5.45

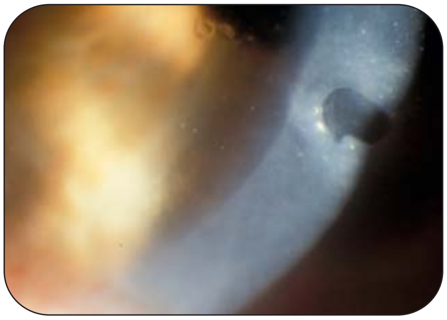

Fig. 5.46

Superior limbic keratoconjunctivitis

Definition

- An uncommon, usually bilateral condition frequently associated with thyroid dysfunction.

Signs

- Superior corneal filaments, bulbar thickening and injection (**Fig. 5.47**).
- Superior tarsal papillary conjunctivitis (see Figs 3.30 and 3.31).

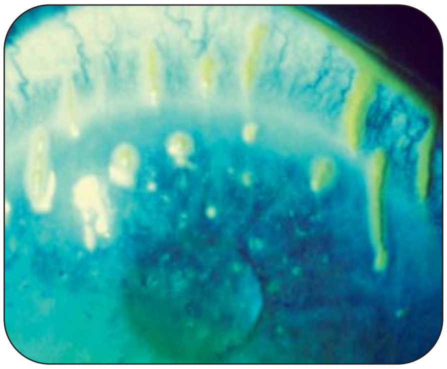

Fig. 5.47

EPITHELIAL DEFECTS AND ULCERS

Dendritic and pseudodendritic ulceration

Herpes simplex dendritic ulcer

Signs

- Single or, less commonly multiple, branching, ulcerating epithelial lesions with raised edges and terminal bulb-like projections.
- The ulcer bed stains with fluorescein (**Fig. 5.48**) and the swollen margins with rose Bengal (**Fig. 5.49**).

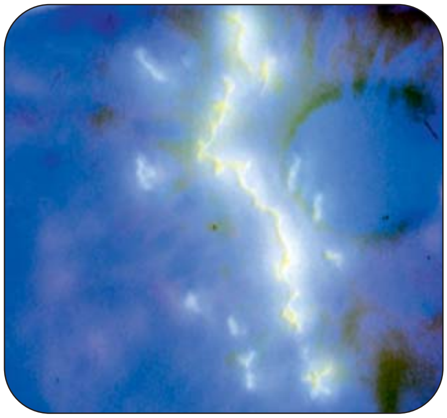

Fig. 5.48

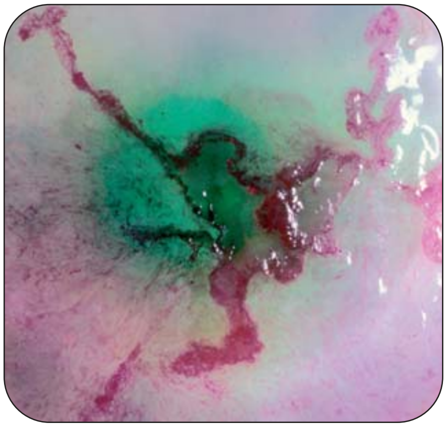

Fig. 5.49

Herpes zoster pseudodendrite

Signs

- Elevated epithelial lesion with tapered ends which lack terminal bulbs and ulceration (**Fig. 5.50**).
- Stains mildly with fluorescein.

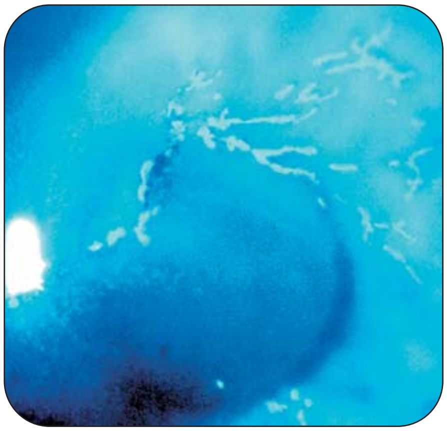

Fig. 5.50

Acanthamoeba pseudodendrite

Signs

- A pseudodendrite (**Fig. 5.51**) is an early sign of acanthamoeba infection which typically affects contact lens wearers.

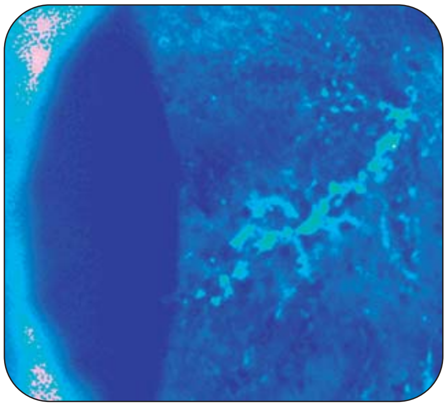

Fig. 5.51

Other causes of pseudodendrites

a. *Epithelial rejection line* in a corneal graft (**Fig. 5.52**).

b. *Tyrosinaemia type II* may be associated with bilateral central ulceration with coarse branching figures that develop during the first few months of life.

c. *Healing epithelial defects.*

d. *Chronic epithelial toxicity.*

e. *Use of soft contact lenses.*

f. *Latanoprost.*

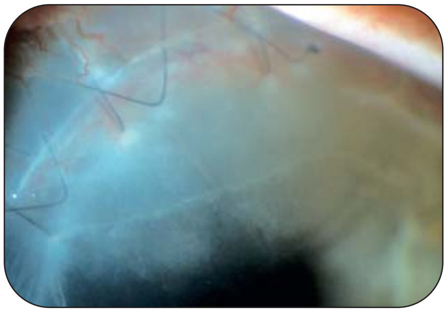

Fig. 5.52

Large epithelial defects

Corneal abrasion

Signs

- Round or oval defect that stains with fluorescein (**Fig. 5.53**).

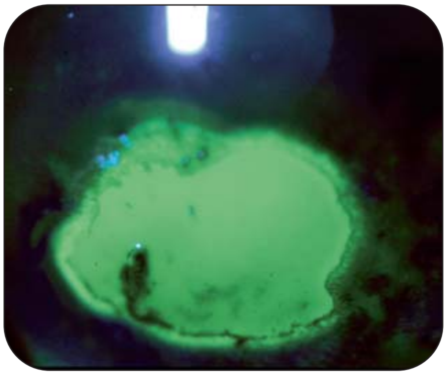

Fig. 5.53

Herpes simplex geographic ulcer

Signs

- Amoeboid-like, fluorescein-staining epithelial defect (**Fig. 5.54**).
- The margin stains with rose Bengal (**Fig. 5.55**).

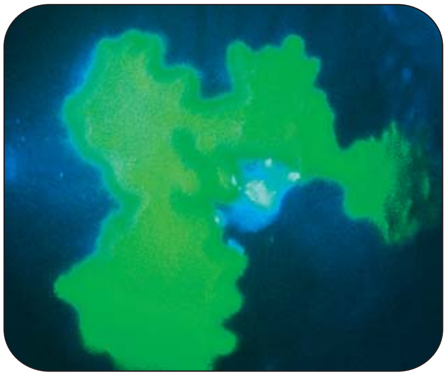

Fig. 5.54

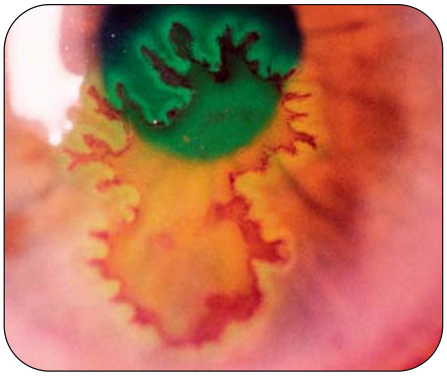

Fig. 5.55

Neurotrophic keratopathy

Cause

- Diminished corneal sensation (Table 5.10).

Signs

- Starts with interpalpebral PEE (see Fig. 5.40).
- Followed by a central epithelial defect which is slow to heal (**Fig. 5.56**).
- Stromal melting and perforation are rare (**Fig. 5.57**).

Table 5.10 **Causes of diminished corneal sensation**

1. **Acquired**
    - Previous corneal disease such as herpes simplex and herpes zoster
    - Damage to the trigeminal nerve or its ganglion
    - Diabetes
    - Leprosy
2. **Congenital**
    - Riley–Day syndrome (familial dysautonomia)
    - Möbius syndrome
    - Goldenhar syndrome
    - Anhidrotic ectodermal dysplasia
    - Hereditary sensory neuropathy

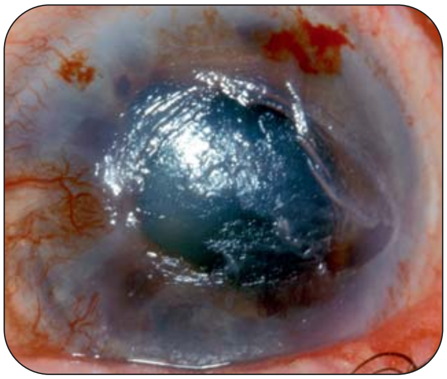

Fig. 5.56

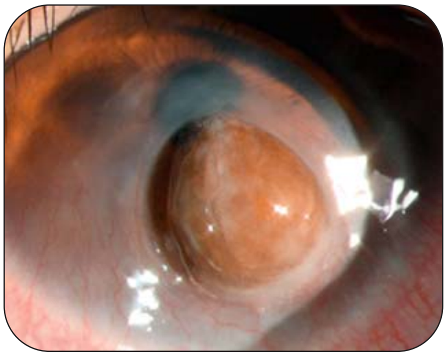

Fig. 5.57

Exposure keratopathy

Cause

- Incomplete lid closure (lagophthalmos).

Signs

- Starts with inferior PEE.
- Followed by an inferior epithelial defect (**Fig. 5.58**).
- Stromal melting (**Fig. 5.59**) and perforation are rare.

Causes

- Facial nerve palsy (**Fig. 5.60**).
- Eyelid scarring or tight facial skin that prevents lid closure (**Fig. 5.61**).
- Severe proptosis (**Fig. 5.62**).
- Reduced muscle tone – as in coma and Parkinsonism.

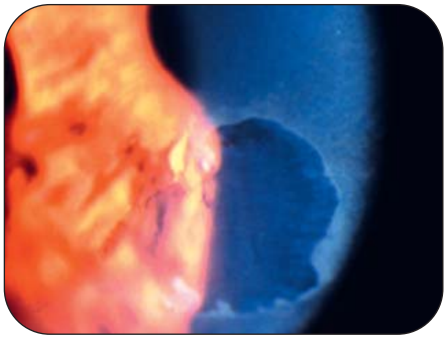
Fig. 5.58

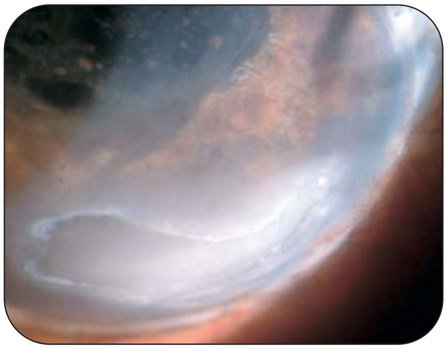
Fig. 5.59

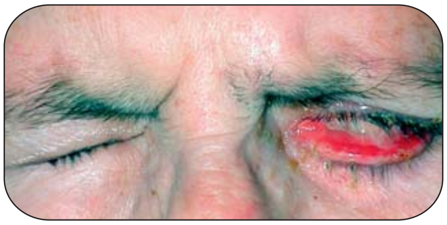
Fig. 5.60

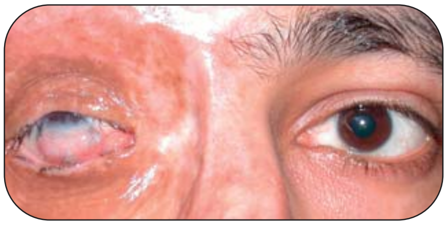
Fig. 5.61

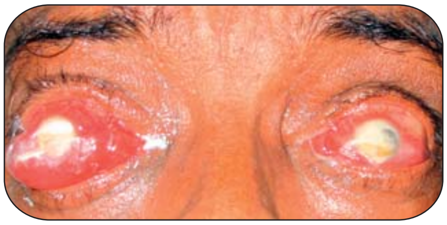
Fig. 5.62

Epithelial macroerosion

Cause

- Vernal disease or atopic keratoconjunctivitis.

Signs

- Superior, oval or pentagonal defect that stains with fluorescein (**Fig. 5.63**).
- May be associated with greyish opacification of the bed and slightly elevated margins.

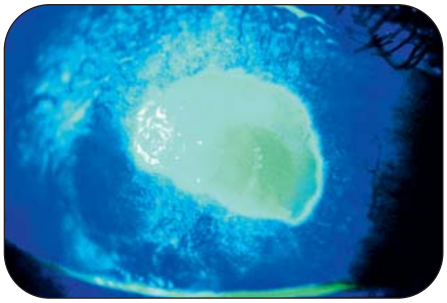
Fig. 5.63

Microbial ulcers

Bacterial keratitis

Definition

- A serious unilateral condition which develops in persons with pre-existing ocular surface disease or in those who wear contact lenses.

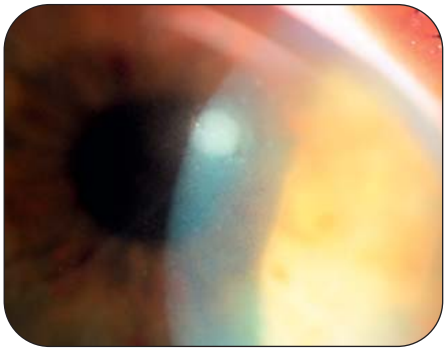

Fig. 5.64

Signs

- White stromal infiltrate associated with an overlying epithelial defect (**Fig. 5.64**) and secondary anterior uveitis.
- Enlargement of stromal infiltration associated with stromal oedema (**Fig. 5.65**).
- Severe infiltration and hypopyon formation (**Fig. 5.66**).
- Progressive ulceration and enlargement of hypopyon (**Fig. 5.67**).
- Corneal perforation and endophthalmitis in neglected cases (see Fig. 5.62).

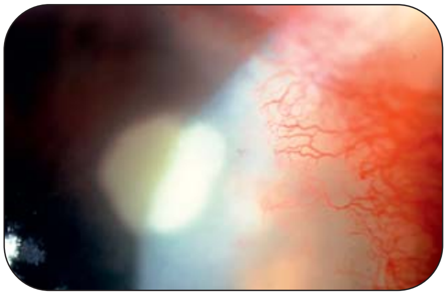

Fig. 5.65

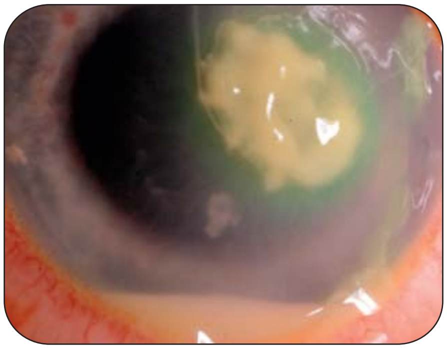

Fig. 5.66

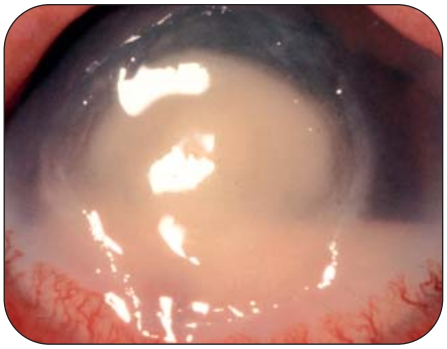

Fig. 5.67

Candida keratitis

Definition

- A rare, unilateral, insidious fungal infection which usually occurs in eyes with pre-existing chronic corneal disease or in severely debilitated patients.

Signs

- Grey-white infiltrate (**Fig. 5.68**) similar to a bacterial ulcer.

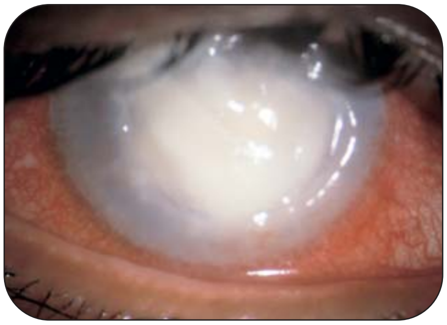

Fig. 5.68

Filamentous keratitis

Definition

- A rare, unilateral, insidious fungal infection which is typically preceded by ocular trauma with wood.

Signs

- Greyish-white infiltrate with indistinct feathery margins.
- Often surrounded by finger-like satellite stromal infiltrates (**Fig. 5.69**).

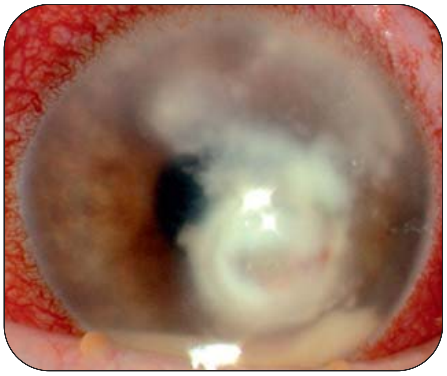

Fig. 5.69

Acanthamoeba keratitis

Definition

- A very rare, unilateral condition which typically affects wearers of soft contact lenses.

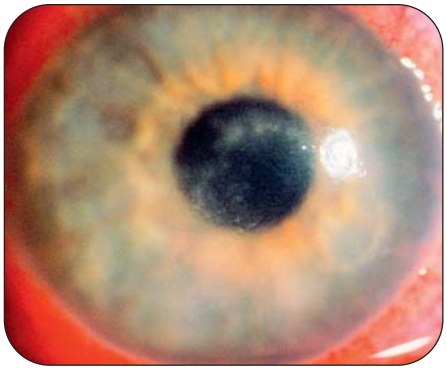

Fig. 5.70

Signs

- Epithelial pseudodendrite (see Fig. 5.51).
- Non-specific stromal infiltrates (**Fig. 5.70**).
- Radial keratoneuritis (**Fig. 5.71**).
- Ring infiltrate (**Fig. 5.72**).
- Corneal necrosis and thinning (**Fig. 5.73**) in advanced cases.

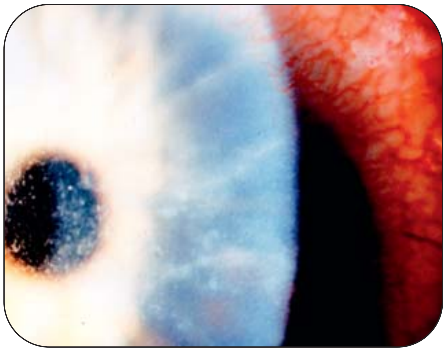

Fig. 5.71

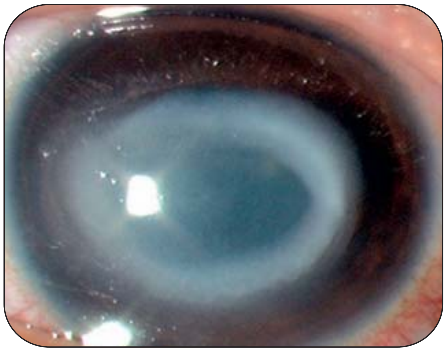

Fig. 5.72

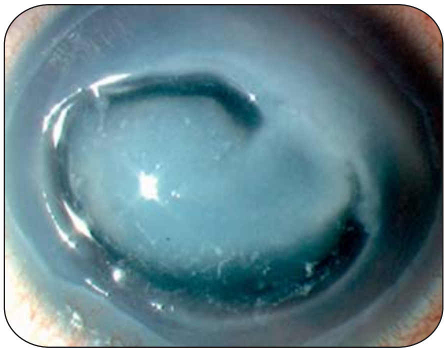

Fig. 5.73

EPITHELIAL DEPOSITS

Vortex keratopathy

Definition

- A bilateral condition which occurs in patients with Fabry disease but is more commonly caused by a variety of drugs (Table 5.11).

Signs

- Greyish or golden epithelial deposits arranged in a vortex fashion from a point below the pupil and swirling outward but sparing the limbus (**Fig. 5.74**).

Table 5.11 **Drug causes of vortex keratopathy**

- Chloroquine
- Hydroxychloroquine
- Amiodarone
- Indomethacin
- Tamoxifen
- Chlorpromazine
- Atovaquone

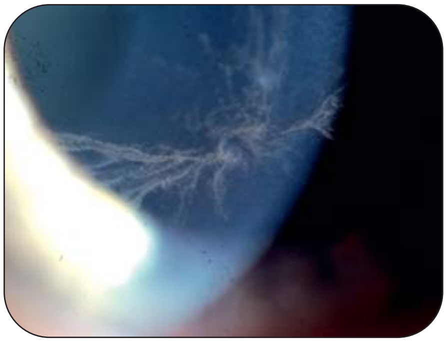

Fig. 5.74

Adrenochrome

Cause

- Topical adrenaline drops for glaucoma may cause black deposits in the conjunctiva (see Fig. 3.67) and rarely on the corneal surface (**Fig. 5.75**).

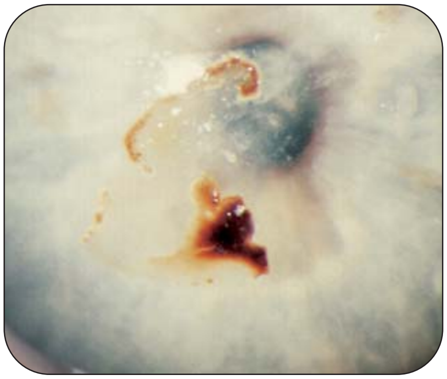

Fig. 5.75

Fluoroquinolones

Cause

- Topical ciprofloxacin and norfloxacin used to treat bacterial keratitis may become deposited within an epithelial defect (**Fig. 5.76**).

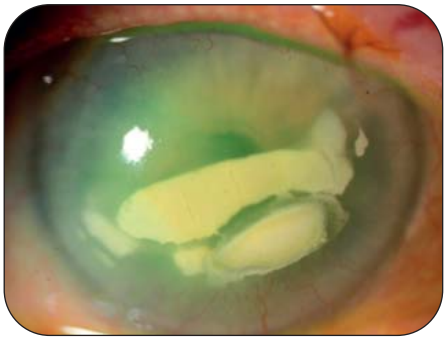

Fig. 5.76

Epithelial iron lines

a. *Rust ring* consists of residual rust following the removal of a metallic foreign body.

b. *Hudson–Stähli* line occurs at the junction of the upper two-thirds and the lower third of an otherwise normal cornea.

c. *Ferry line* occurs in front of a filtering bleb.

d. *Stocker line* occurs in front of a pterygium.

e. *Fleischer ring* occurs at the base of the cone in keratoconus.

f. *Dalgleisch line* is found in the inferior cornea in patients with hereditary spherocytosis.

g. *Other* iron lines may be found adjacent to corneal elevations in Salzmann degeneration, corneal grafts and radial keratotomy incisions.

DEGENERATIONS IN BOWMAN LAYER AND ANTERIOR STROMA

Band keratopathy

Definition

- A common condition which may be bilateral or unilateral depending on the cause (Table 5.12).

Signs

- Peripheral interpalpebral calcification with clear cornea separating the sharp peripheral margins of the band from the limbus (**Fig. 5.77**).
- Gradual central spread that acquires a ground-glass appearance that may contain small holes (**Fig. 5.78**).

Table 5.12 **Causes of band keratopathy**

1. **Idiopathic age-related**
2. **Ocular**
    - Chronic iridocyclitis, particularly in children
    - Phthisis bulbi (**Fig. 5.79**)
    - Silicone oil in the anterior chamber (**Fig. 5.80**)
3. **Metabolic**
    - Increased serum calcium and phosphorus
    - Gout and other causes of hyperuricaemia
    - Chronic renal failure
4. **Hereditary**
    - Familial
    - Hereditary ichthyosis

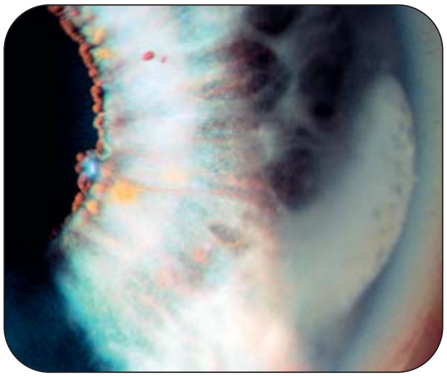

Fig. 5.77

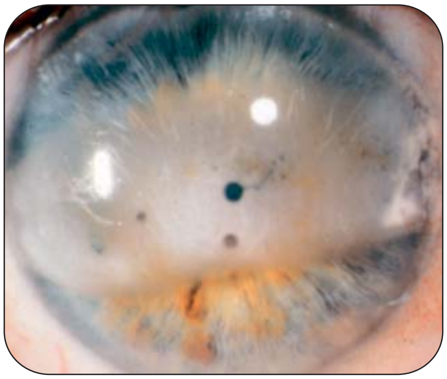

Fig. 5.78

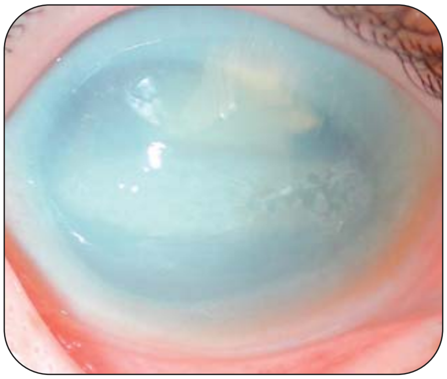

Fig. 5.79

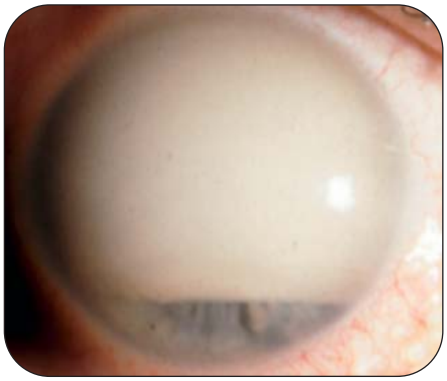

Fig. 5.80

Spheroidal degeneration

Definition

- A rare, bilateral condition which typically affects outdoor workers although in some cases it is secondary to other corneal diseases.

Signs

- Interpalpebral, small amber-coloured granules in the superficial stroma and conjunctiva.
- Gradual central spread, coalescence and opacification (**Fig. 5.81**).

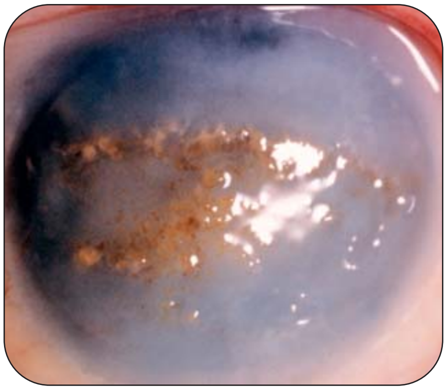

Fig. 5.81

Lipid keratopathy

Definition

- A unilateral condition that is occasionally idiopathic (primary) but most frequently is associated with previous herpes simplex or herpes zoster keratitis (secondary).

Signs

- Primary lesions are avascular yellowish deposit with feathery edges that may rarely involve the entire cornea (**Fig. 5.82**).
- Secondary lesions are associated with vascularisation (**Fig. 5.83**).

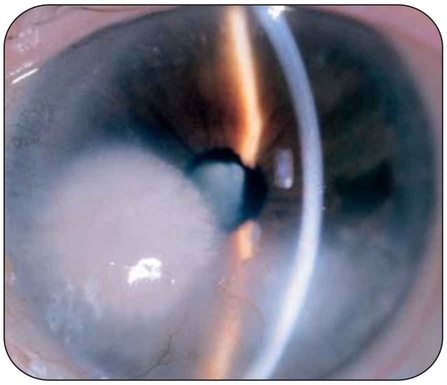

Fig. 5.82

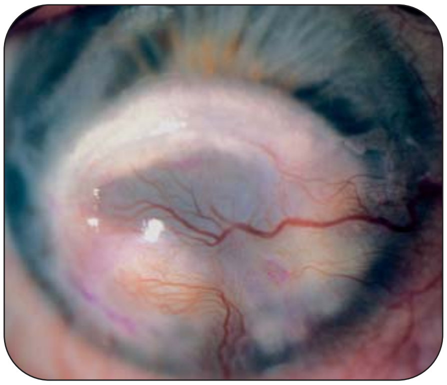

Fig. 5.83

Salzmann nodular degeneration

Definition

- A usually unilateral condition which is always secondary to a chronic corneal disease.

Signs

- Discrete, elevated, grey, superficial stromal nodules which may be surrounded by iron-pigment deposits (**Fig. 5.84**).

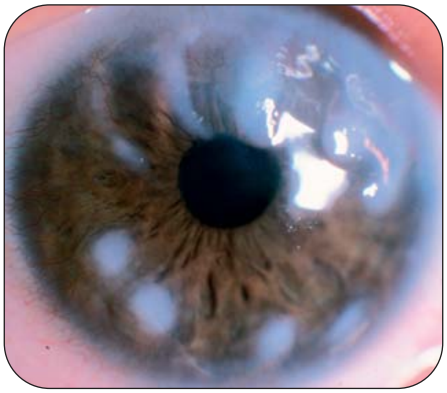

Fig. 5.84

Infectious crystalline keratopathy

Definition

- An uncommon unilateral condition that may develop as a complication of penetrating keratoplasty and the long-term use of topical steroids.

Signs

- Discrete, white, branching anterior stromal crystalline deposits without associated inflammation (**Fig. 5.85**).

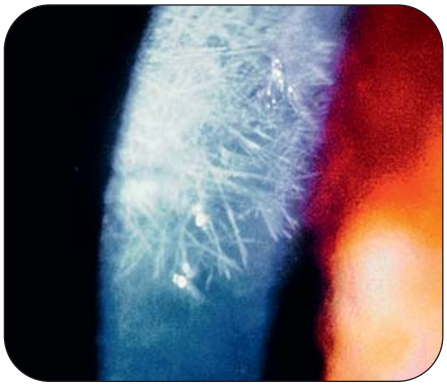

Fig. 5.85

DYSTROPHIES OF BOWMAN LAYER AND ANTERIOR STROMA

Reis–Bückler dystrophy (Bowman layer 1)

Definition

- An AD condition that presents in the 1st or 2nd decade with severe recurrent corneal erosions.

Signs

- Fine, grey-white, round and polygonal opacities in Bowman layer (**Fig. 5.86**).

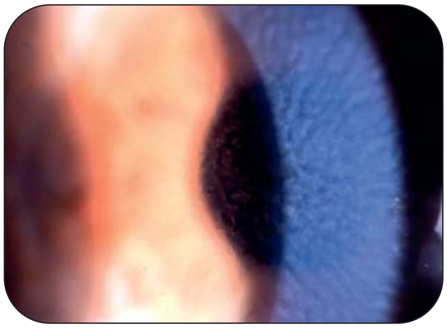

Fig. 5.86

Thiel–Behnke dystrophy (Bowman layer 2)

Definition

- An AD condition that presents at the end of the 1st decade with recurrent erosions.

Signs

- Similar to Reis–Bückler dystrophy except that the opacities have a more honeycomb pattern (**Fig. 5.87**).

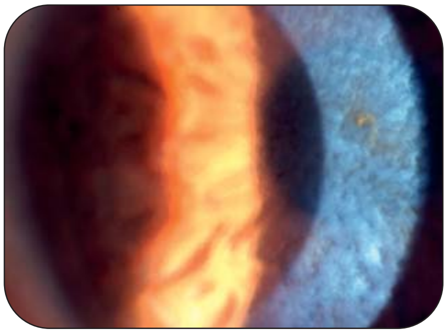

Fig. 5.87

Central Schnyder (crystalline) dystrophy

Definition

- An AD condition that presents in the 2nd decade with visual impairment and glare.

Signs

- Central, oval, subepithelial ‘crystalline’ opacity composed of phospholipids and cholesterol (**Fig. 5.88**).

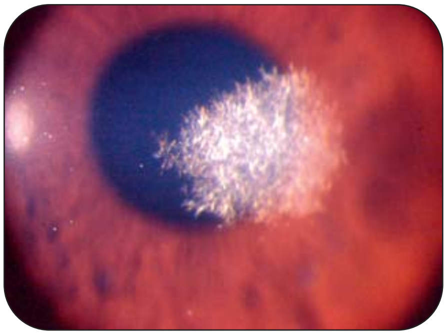

Fig. 5.88

Gelatinous drop-like dystrophy

Definition

- An AR condition that presents in the 1st to 2nd decades with severe photophobia, watering and visual impairment.

Signs

- Grey subepithelial nodules consisting of amyloid that gradually become confluent and involve the stroma giving rise to a mulberry-like appearance (**Fig. 5.89**).

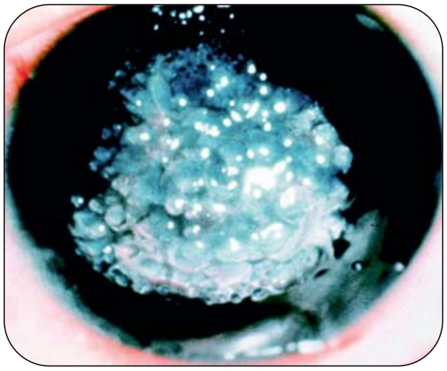

Fig. 5.89

DEPOSITS IN STROMA AND DESCEMET MEMBRANE

Metabolic

Cystinosis

Definition

- An AR condition characterised by widespread tissue deposition of cystine crystals.

Signs

- Initially iridescent crystals in the anterior peripheral stroma.
- Subsequent deep and central involvement (**Fig. 5.90**).

Look for

- Retinal deposits which are rare.

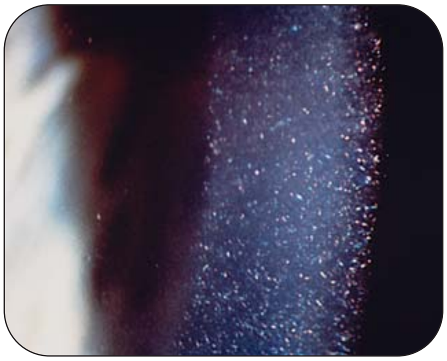

Fig. 5.90

Kayser–Fleischer ring

Definition

- A rare, bilateral condition that occurs primarily in patients with Wilson disease and occasionally in association with other liver disorders, multiple myeloma and carotenaemia.

Signs

- Circumferential, greenish-brown, peripheral band 1–3 mm in width located at the level of Descemet membrane (**Fig. 5.91**).

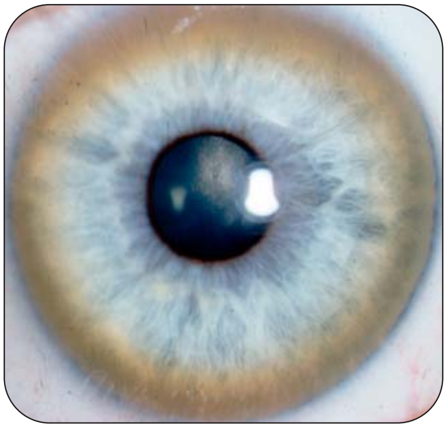

Fig. 5.91

Immunoprotein deposits

Signs

- Refractile or polychromatic crystals usually involve the entire stroma (**Fig. 5.92**).

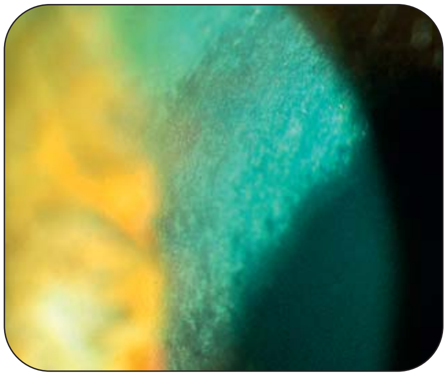

Fig. 5.92

Causes

- Primary amyloidosis.
- Multiple myeloma.
- Waldenström macroglobulinaemia.
- Lymphoma.
- Benign monoclonal gammopathy.
- Cryoglobulinaemia.

Mucopolysaccharidoses

Definition

- Lysosomal storage diseases that result from deficiency of enzymes involved in the degradation of glycosaminoglycans.

Classification

a. *Hurler syndrome* (MPS I-H) is associated with significant stromal clouding (see Fig. 5.25) which helps to differentiate it from Hunter syndrome which has similar systemic features but does not manifest corneal changes until later in life.

b. *Scheie syndrome* (MPS I-S) is associated with severe clouding in which the cornea appears thickened and oedematous.

c. *Morquio syndrome* (MPS types IVA and IVB) is associated with variable haziness.

d. *Maroteaux–Lamy syndrome* (types VIA and VIB) is associated with mild punctate opacities.

Hypoproteinaemias

Causes

- Abnormal reductions of serum lipoprotein levels.

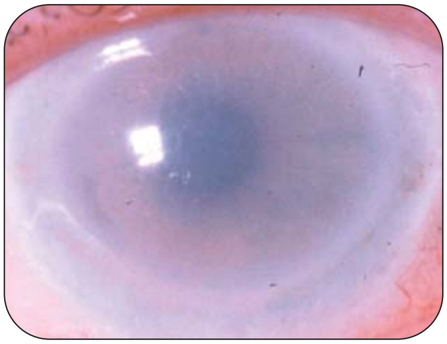

Fig. 5.93

Classification

a. *Familial lecithin-cholesterol-acyltransferase deficiency (Norum disease)* is an AR disease characterised by hyperlipidaemia, early atherosclerosis, anaemia and renal disease. Numerous minute greyish dots are seen throughout the stroma and are often concentrated in the periphery in an arcus-like configuration (**Fig. 5.93**).

b. *Tangier disease* is an AR condition characterised by hepatosplenomegaly, neuropathy, hypertrophic orange-yellow tonsils and lymphadenopathy. Corneal cholesterol deposits cause diffuse clouding and focal deep stromal opacities.

c. *Fish eye disease* is an AR condition characterised by elevated serum cholesterol, triglycerides and very-low-density lipoprotein levels but depressed HDL. Diffuse clouding is caused by minute grey-white opacities that are more concentrated peripherally.

Non-metabolic

Chrysiasis

Definition

- The deposition of gold in tissues as a result of long-term administration of gold in the treatment of rheumatoid arthritis.

Signs

- Dust-like or glittering yellow-brown granules throughout the deep stroma and Descemet membrane (**Fig. 5.94**).

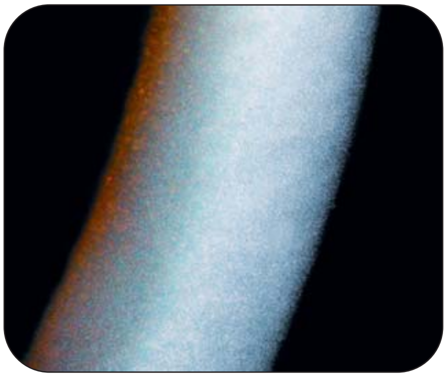

Fig. 5.94

Argyrosis

Definition

- Discoloration of ocular tissues secondary to industrial exposure or topical medication with a silver nitrate compound (now seldom used).

Signs

- Diffuse greyish-brown discoloration of Descemet membrane (**Fig. 5.95**) often associated with conjunctival involvement (see Fig. 3.69).

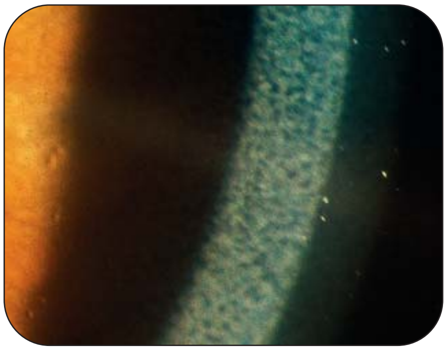

Fig. 5.95

Phenothiazines

Phenothiazines are used as sedatives and to treat psychotic illnesses. Occasionally long-term administration may cause innocuous corneal deposits.

Signs

- Subtle, diffuse, yellowish-brown granular stromal deposits (**Fig. 5.96**).

Look for

- Anterior subcapsular lens deposits (see Fig. 10.47) which may antedate the corneal changes.

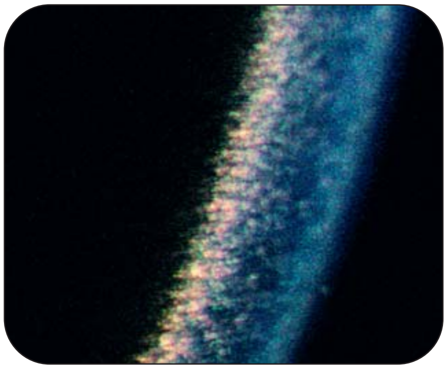

Fig. 5.96

Bietti crystalline dystrophy

Definition

- A rare, innocuous AR condition.

Signs

- Anterior stromal crystals (**Fig. 5.97**).

Look for

- Crystalline retinopathy (see Fig. 13.166).

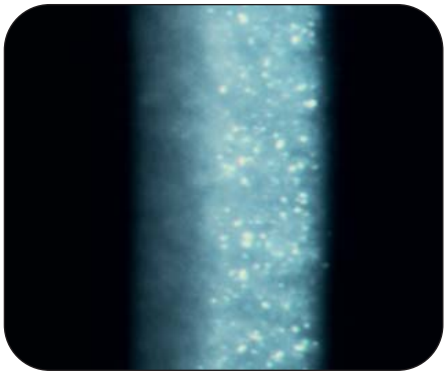

Fig. 5.97

Cornea farinata

Definition

- A sporadic, bilateral, innocuous condition.

Signs

- Minute, subtle, flour-like deposits in the deep stroma, most prominent centrally (**Fig. 5.98**).

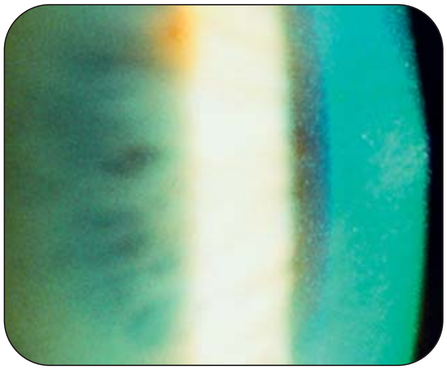

Fig. 5.98

STROMAL DYSTROPHIES

Lattice dystrophy type 1 (Biber–Haab–Dimmer)

Definition

- An AD condition that presents at the end of the 1st decade with recurrent erosions which precede the dystrophic changes.

Signs

- Numerous fine branching subepithelial lattice lines composed of amyloid associated with stromal haze that is most dense centrally but spares the limbus (**Fig. 5.99**).

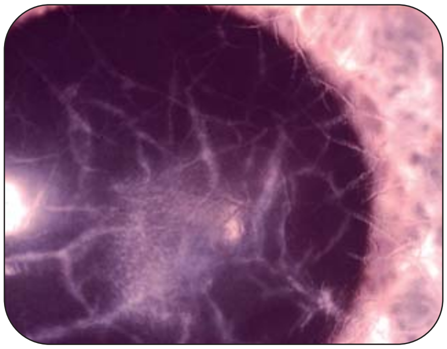

Fig. 5.99

Lattice dystrophy type 2 (Meretoja syndrome)

Definition

- An AD condition that is associated with systemic amyloidosis. Onset is in the 3rd decade but recurrent erosions are uncommon.

Signs

- Lattice lines extend to the limbus and are more delicate and more radial (**Fig. 5.100**) than in lattice dystrophy type 1.

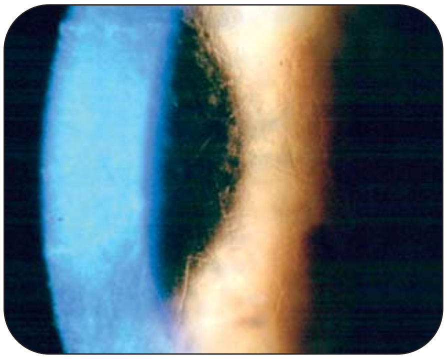

Fig. 5.100

Lattice dystrophy type 3

Definition

- An AR condition that presents between the 4th and 6th decades.

Signs

- Thick ropy lattice lines with minimal intervening haze (**Fig. 5.101**).

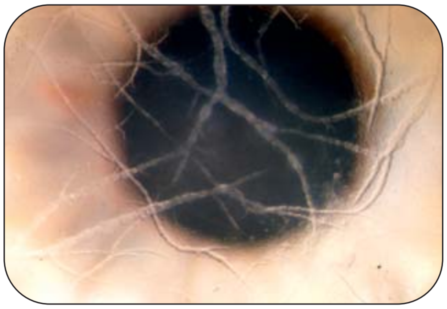

Fig. 5.101

Granular dystrophy type 1

Definition

- An AD condition that begins in the 1st decade but vision is not affected until later.

Signs

- Small, white, central deposits that resemble crumbs, rings or snowflakes (**Fig. 5.102**).

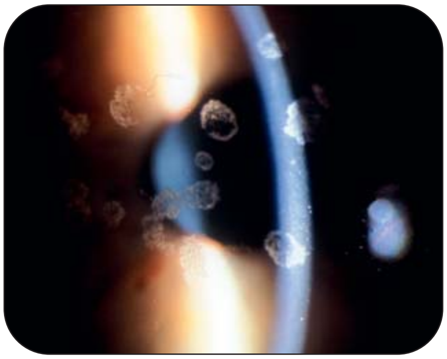

Fig. 5.102

Granular dystrophy type 2 (Avellino)

Definition

- An AD condition that presents in the 2nd decade with recurrent corneal erosions.

Signs

- Initial anterior stroma opacities similar to granular dystrophy type 1.
- Later posterior stromal lattice-like lesions (**Fig. 5.103**).

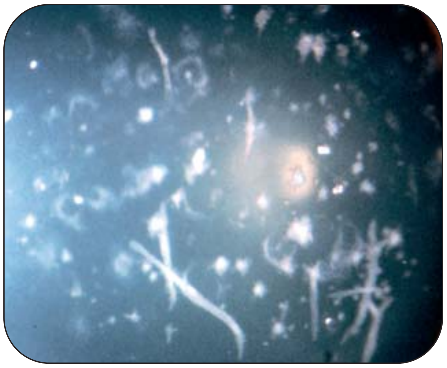

Fig. 5.103

Macular dystrophy

Definition

- An AR condition that starts in the 2nd decade and causes progressive visual deterioration.

Signs

- Initial diffuse superficial central stroma haze.
- Later greyish-white, dense, poorly delineated spots (**Fig. 5.104**).

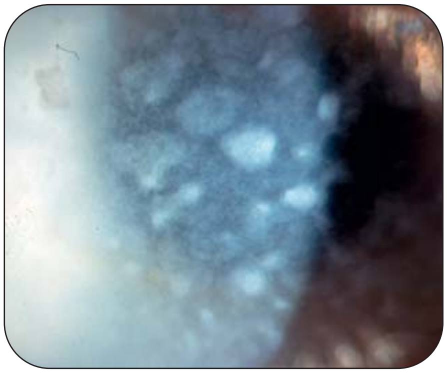

Fig. 5.104

Central cloudy dystrophy of François

Definition

- A rare innocuous AD condition.

Signs

- Polygonal, cloudy grey opacities separated by relatively clear spaces, in the posterior stroma most prominent centrally, creating a leather-like appearance (**Fig. 5.105**).

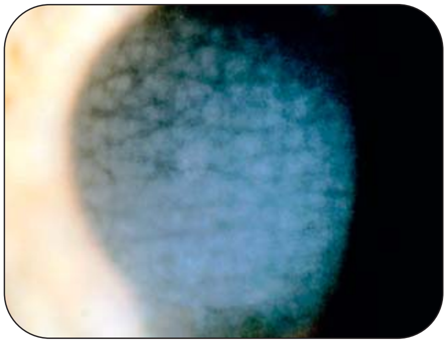

Fig. 5.105

Fleck dystrophy

Definition

- A rare innocuous AD condition.

Signs

- Discrete, grey-white flecks involve the entire stroma.

Table 5.13 **Age of onset of corneal dystrophies**

1. **Perinatal**
   - Congenital hereditary endothelial dystrophy
   - Posterior polymorphous dystrophy
   - Congenital hereditary stromal dystrophy
2. **Early childhood**
   - Meesmann
   - Reis–Bückler dystrophy (Bowman layer 1)
   - Thiel–Behnke dystrophy (Bowman layer 2)
   - Lattice dystrophy type 1 (Biber–Haab–Dimmer)
   - Granular dystrophy type 1
   - Macular dystrophy
   - Gelatinous drop-like dystrophy
3. **Late childhood**
   - Epithelial basement membrane (Cogan) dystrophy
   - Central crystalline (Schnyder) dystrophy
   - Granular dystrophy type 2 (Avellino)
4. **Adulthood**
   - Lattice dystrophy type 2 (Meretoja syndrome)
   - Lattice dystrophy type 3
   - Fuchs endothelial dystrophy

INTERSTITIAL KERATITIS

Signs

- Bilateral, mid-stromal, vascularised (**Fig. 5.106**) and non-suppurative infiltration.
- Inactive signs include stromal scarring (**Fig. 5.107**) associated with non-perfused (ghost) vessels (**Fig. 5.108**).
- Rarely, reperfusion may give rise to an intrastromal haemorrhage (**Fig. 5.109**).

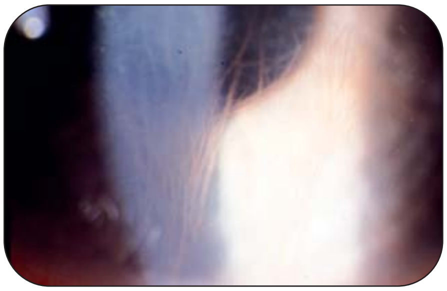

Fig. 5.106

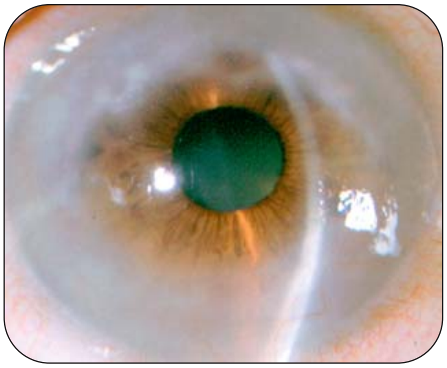

Fig. 5.107

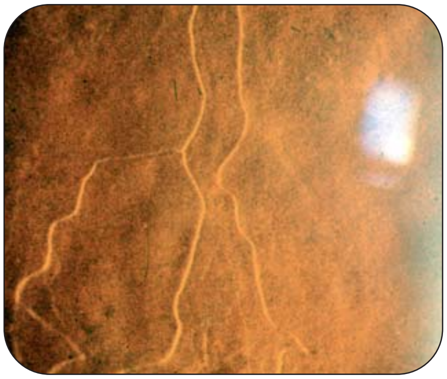

Fig. 5.108

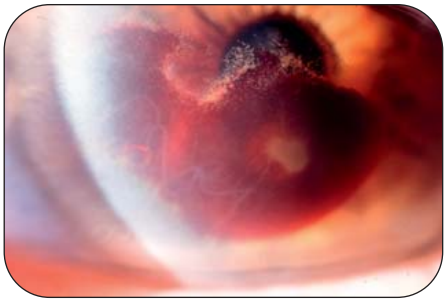

Fig. 5.109

Systemic associations

Congenital syphilis

Look for

- Sensorineural deafness and saddle-shaped nasal deformity (**Fig. 5.110**).
- Dental anomalies including malformed incisors (Hutchinson teeth) and mulberry molars.

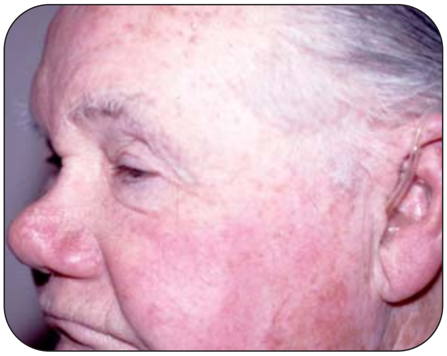

Fig. 5.110

Acquired syphilis

Look for

- Maculopapular rash on the trunk, soles and palms (**Fig. 5.111**).
- Fever, malaise and generalised lymphadenopathy.

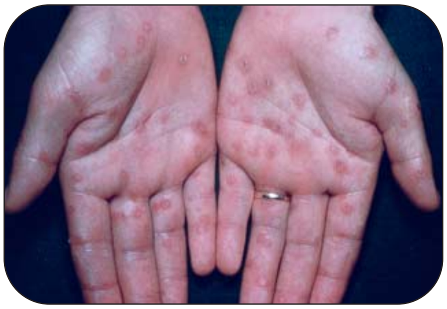

Fig. 5.111

Lyme disease (stage 3)

Look for

- Arthritis.
- Demyelinating encephalopathy.

Lymphogranuloma venereum

Look for

- Painless genital ulceration.
- Painful regional lymphadenopathy (**Fig. 5.112**).

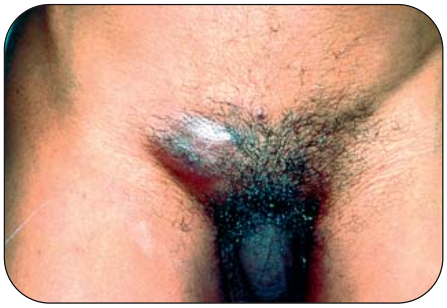

Fig. 5.112

Cogan syndrome

Look for

- Acute-onset tinnitus, vertigo and deafness.

Miscellaneous

a. *Tuberculosis.*

b. *Leprosy.*

c. *Epstein–Barr.*

d. *Mumps.*

e. *Onchocerciasis.*

PERIPHERAL LESIONS

Circumcorneal opacification

Vogt white limbal girdle

Definition

- A very common, bilateral, innocuous, age-related condition.

Signs

- Crescentic, chalky white, linear opacities in the interpalpebral fissures along the nasal and temporal limbus (**Fig. 5.113**).

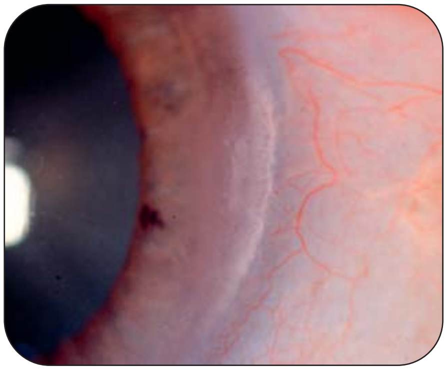

Fig. 5.113

Corneal arcus

Definition

- A very common, bilateral condition which may be age-related (arcus senilis) or associated with types II and III hyperlipidaemia in younger individuals (arcus lipoides).

Signs

- White limbal band 1–2 mm in diameter with a sharp outline and a more diffuse central boundary located in the superficial stoma (**Fig. 5.114**).

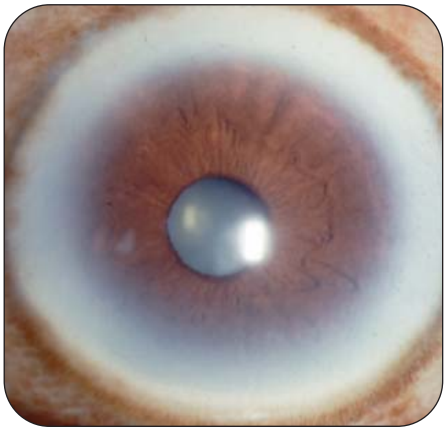

Fig. 5.114

Pseudogerontoxon (anterior embryotoxon)

Definition

- An uncommon, bilateral or unilateral condition which occurs at the site of previous inflammation, most frequently associated with vernal disease.

Signs

- Superior corneal opacity with a cupid's-bow configuration (**Fig. 5.115**).

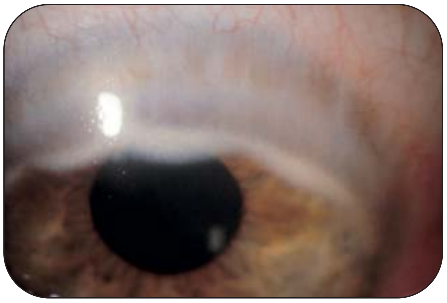

Fig. 5.115

Posterior embryotoxon

Definition

- A bilateral condition which occurs to some extent in about 15% of normals, in 80% of patients with Alagille syndrome and in all patients with Axenfeld–Rieger anomaly (see Fig. 8.89).

Signs

- Thickened and anteriorly displaced Schwalbe line (**Fig. 5.116**).

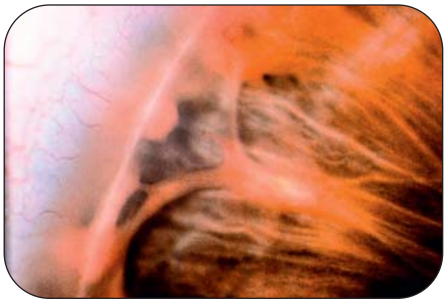

Fig. 5.116

Kayser–Fleischer ring

- See Fig. 5.91.

Infiltrates

Marginal keratitis

Definition

- A very common, innocuous, transient, usually unilateral condition that may be associated with staphylococcal blepharitis.

Signs

- White subepithelial infiltrate separated from the limbus by clear cornea (**Fig. 5.117**).
- May be followed by epithelial breakdown and circumferential spread (**Fig. 5.118**).

Differential diagnosis

- Bacterial keratitis (Table 5.14).

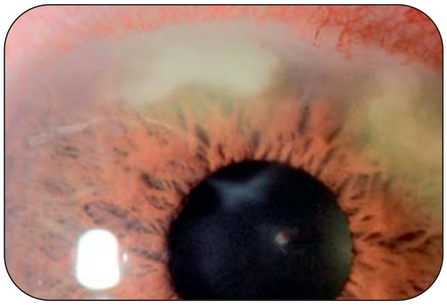
Fig. 5.117

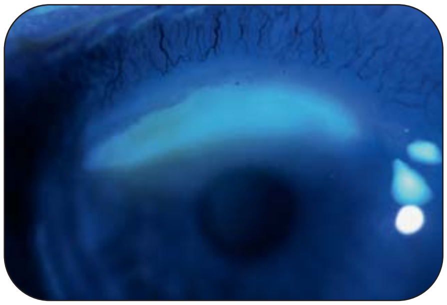
Fig. 5.118

Table 5.14 **Comparison between marginal keratitis and bacterial keratitis**

	Marginal keratitis	**Bacterial keratitis**
Location	Peripheral	Central
Size	<1 mm	>1 mm
Epithelial defect	Small or absent	Present
Uveitis	Absent	Present

Rosacea keratitis

Definition

- An uncommon bilateral, chronic condition which affects approximately 5% of patients with acne rosacea.

Signs

- Inferior vascularisation associated with subepithelial infiltration (**Fig. 5.119**).
- May be followed by stromal thinning (**Fig. 5.120**).

Look for

- Erythema and telangiectasia involving the glabella, cheeks, nose and chin (see Fig. 3.47).

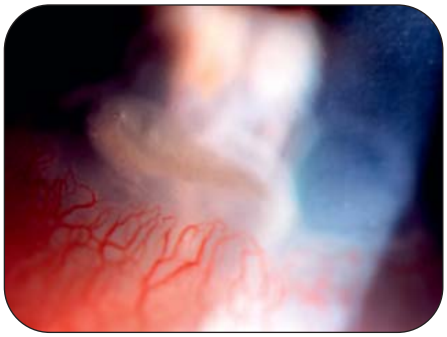
Fig. 5.119

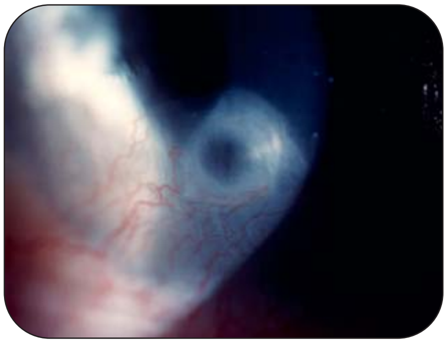
Fig. 5.120

Chlamydial keratitis

Definition

- An uncommon, usually bilateral, chronic condition.

Signs

- Chronic follicular conjunctivitis (see Fig. 3.22).
- Multiple, small, peripheral infiltrates usually involving the superior cornea (**Fig. 5.121**).

Look for

- Non-specific urethritis in men, and abacterial pyuria and cervicitis in women.

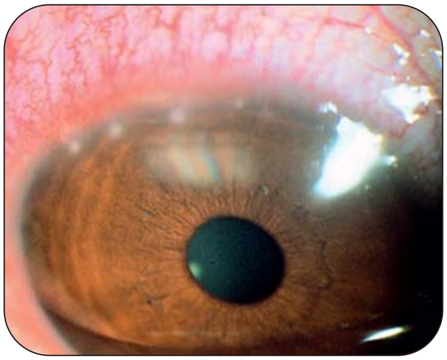

Fig. 5.121

Phlyctenulosis

Definition

- An uncommon, usually unilateral condition that affects children.

Signs

- Small pinkish-white limbal nodule that may spread centrally onto the cornea (**Fig. 5.122**).

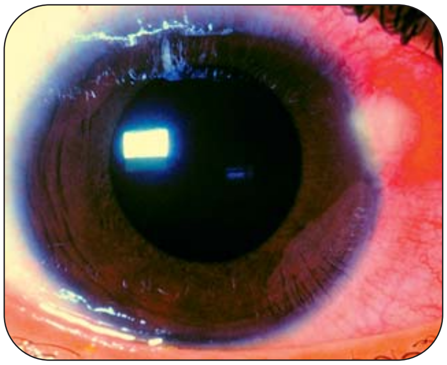

Fig. 5.122

Reiter syndrome

Signs

- Subepithelial opacities may occur in isolation or associated with conjunctivitis.

Look for

- Urethritis.
- Arthritis.
- Circinate balanitis (see Fig. 15.102).
- Keratoderma blenorrhagica (see Fig. 3.40).

Exposure keratopathy

Exposure keratopathy (see above) may be associated with sterile infiltrates similar to those found in marginal keratitis but only involving the inferior cornea.

Sclerosing keratitis

Definition

- A rare, chronic condition which may occur either in isolation or adjacent to scleritis.

Signs

- Gradual peripheral stromal thickening and opacification which may be associated with subsequent vascularisation and lipid deposition (**Fig. 5.123**).

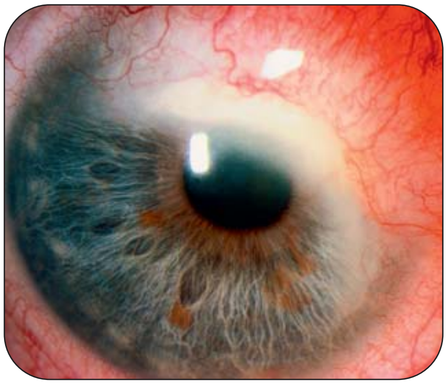

Fig. 5.123

Acute stromal keratitis

Definition

- A rare unilateral condition that typically occurs in patients with rheumatoid arthritis.

Signs

- Peripheral stromal infiltration that coalesces and spreads circumferentially (**Fig. 5.124**).
- The overlying epithelium may break down resulting in peripheral ulcerative keratitis (see Fig. 5.131).

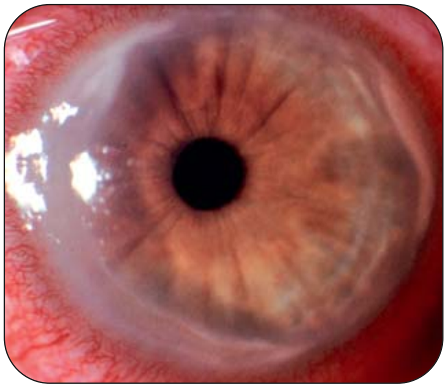

Fig. 5.124

Inflammatory bowel disease

Crohn disease and ulcerative colitis are occasionally associated with peripheral infiltrates.

Thinning and ulceration

Furrow degeneration

Definition

- A very common, bilateral, innocuous, age-related condition.

Signs

- Mild thinning peripheral to corneal arcus (**Fig. 5.125**).

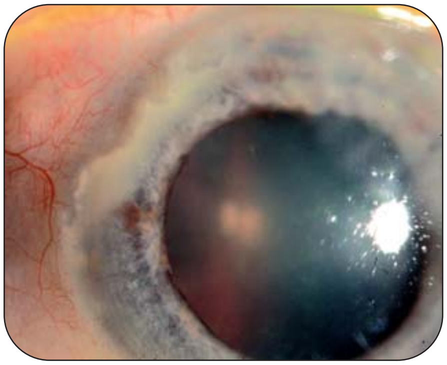

Fig. 5.125

Dellen

Definition

- A common, unilateral, innocuous condition.

Signs

- Saucer-like thinning secondary to local stromal dehydration with intact epithelium (**Fig. 5.126**).

Look for

- Raised limbal lesions.
- Hard contact lens wear.
- Lid abnormalities and abnormal blinking.

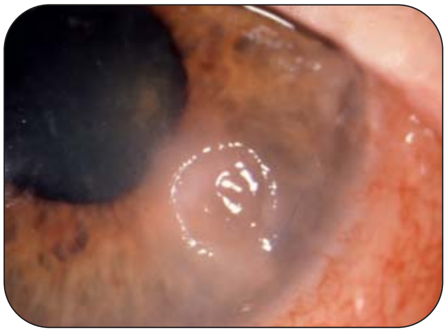
Fig. 5.126

Terrien marginal degeneration

Definition

- An uncommon, usually bilateral, painless condition.

Signs

- Very gradual peripheral thinning associated with superficial vascularisation and lipid deposits central to the thinned edge (**Fig. 5.127**).
- Pseudo-pterygium formation (**Fig. 5.128**) and perforation in long-standing cases.

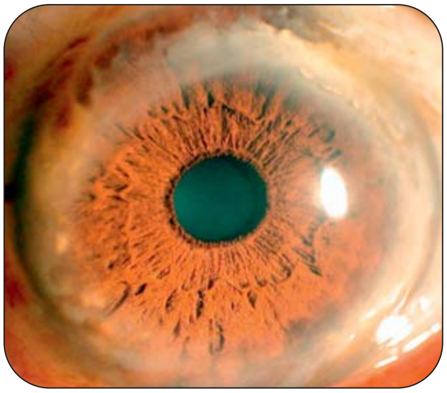
Fig. 5.127A

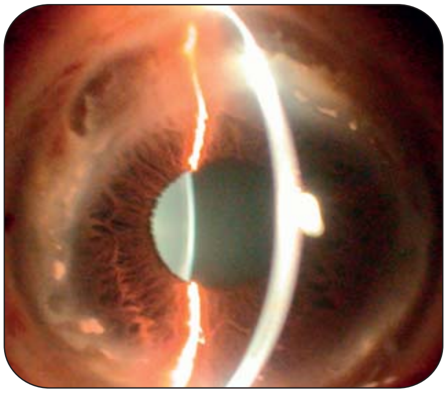
Fig. 5.127B

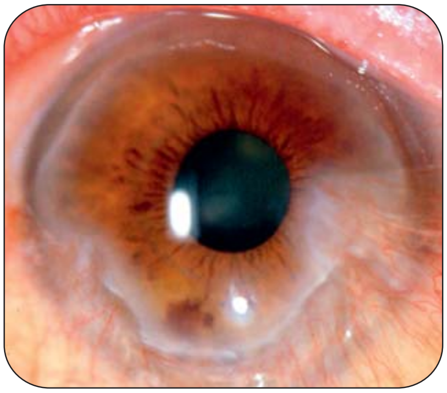
Fig. 5.128

Mooren ulcer

Definition

- A very rare, unilateral or bilateral, painful condition.

Signs

- Peripheral ulceration, with a steep, deeply undermined and infiltrated central edge (**Fig. 5.129**).
- The ulcer may spread centrally and circumferentially but does not invade the sclera (**Fig. 5.130**).

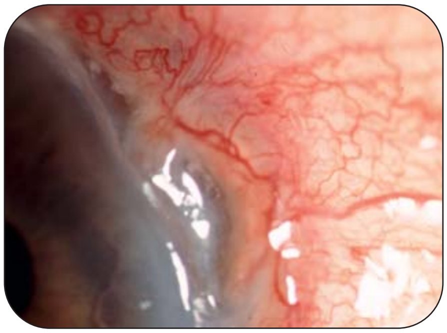
Fig. 5.129

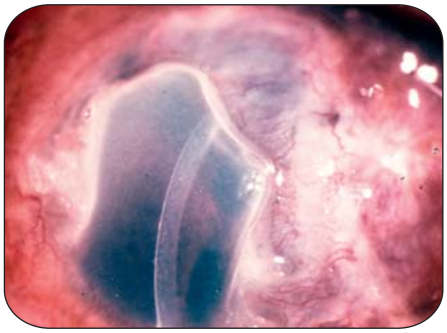
Fig. 5.130

Rheumatoid arthritis

Rheumatoid arthritis is the most common systemic association that may manifest the following lesions which may or may not be associated with adjacent scleritis:

a. *Peripheral ulcerative keratitis* (PUK) characterised by crescent-shaped limbal ulceration with stromal infiltration that spreads circumferentially and centrally and may also involve the sclera (**Fig. 5.131**); perforation may occur in advanced cases.

b. *Asymptomatic guttering* in which there is peripheral stromal thinning with intact epithelium unassociated with infiltration (contact lens cornea – **Fig. 5.132**); perforation may occur in severe cases.

c. *Keratolysis* is characterised by acute usually inferior melting (**Fig. 5.133**) that can lead to perforation (**Fig. 5.134**).

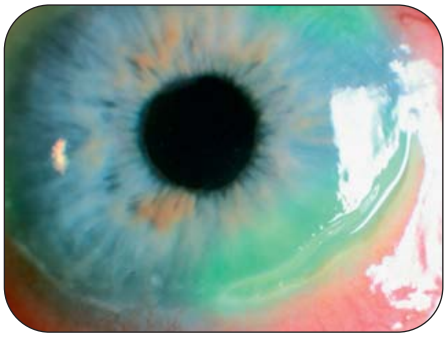

Fig. 5.131

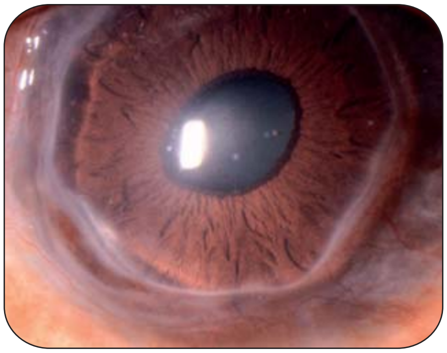

Fig. 5.132

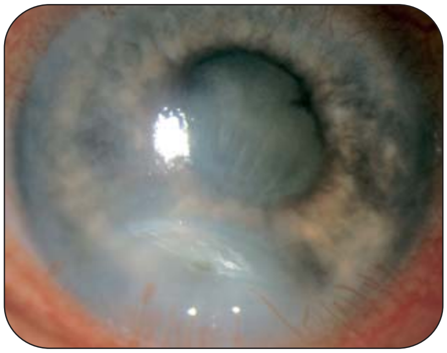

Fig. 5.133

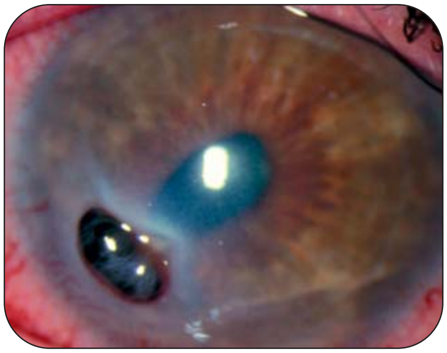

Fig. 5.134

Other diseases

a. *Wegener granulomatosis.*

b. *Polyarteritis nodosa.*

c. *Relapsing polychondritis.*

d. *Systemic lupus erythematosus.*

e. *Inflammatory bowel disease.*

f. *Leukaemia.*

g. *Vitamin A deficiency.*

Anterior chamber 6

ABNORMAL DEPTH 143

Very deep 143

Shallow 143

Shallow chamber following filtration surgery 144

Leaking bleb 144

Excessive filtration 145

Pupil block 145

Malignant glaucoma 145

INFLAMMATORY SIGNS 145

Anterior uveitis 145

Aqueous cells and flare 145

Fibrinous exudate 146

Hypopyon 146

Endophthalmitis 146

Acute postoperative 146

Chronic postoperative 147

Delayed-onset 147

Endogenous infectious 147

NON-INFLAMMATORY CELLS 148

Hyphaema 148

Blunt ocular trauma 148

Post-surgical 148

Causes of spontaneous hyphaema 148

Pseudo-hypopyon 149

Leukaemia cells 149

Retinoblastoma 149

Phacolytic glaucoma 149

Emulsified silicone oil 150

ABNORMAL DEPTH

Very deep

Table 6.1 **Causes of very deep anterior chamber**

1. **Large globe**
   - High myopia (see Fig. 5.8)
   - Buphthalmos (see Fig. 5.7)
2. **Large cornea**
   - Advanced keratoconus (see Fig. 5.9)
   - Keratoglobus (see Fig. 5.14)
   - Megalocornea (**Fig. 6.1**)
3. **Abnormal lens**
   - Aphakia
   - Ectopia lentis
4. **Post-traumatic**
   - Angle recession
   - Posterior rupture of globe
5. **Pigment dispersion syndrome**

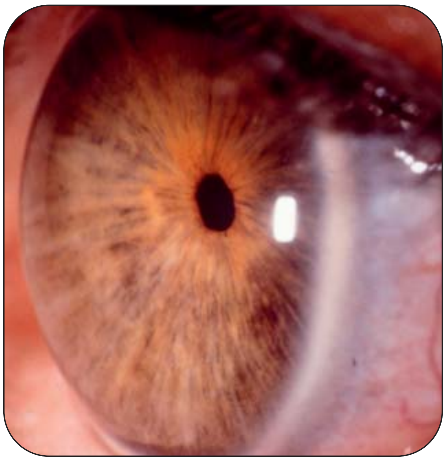

Fig. 6.1

Shallow

Table 6.2 **Causes of shallow anterior chamber**

1. **Small globe**
   - Hypermetropia
   - Nanophthalmos
   - Microphthalmos (see Fig. 5.4)
2. **Small cornea**
   - Microcornea (see Fig. 5.5)
   - Cornea plana (see Fig. 5.17)
3. **Lens-related**
   - Swollen lens
   - Lens dislocated into anterior chamber (**Fig. 6.2**)
4. **Post-traumatic**
   - External leak (**Fig. 6.3**)
   - Cyclodialysis cleft
5. **Postsurgical**
   - Leaking cataract section
   - Pupil block by an anterior chamber lens (**Fig. 6.4**)
   - Expansile gas in vitreous cavity
   - Ciliochoroidal detachments following panretinal photocoagulation
   - Glaucoma filtration surgery (**Fig. 6.5**)
6. **Glaucoma**
   - Acute angle-closure (**Fig. 6.6**)
   - Inflammatory pupil block and iris bombé (**Fig. 6.7**)
7. **Miscellaneous**
   - Ciliary-body tumours and cysts
   - Peters anomaly (**Fig. 6.8**)
   - Iridoschisis (**Fig. 6.9**)

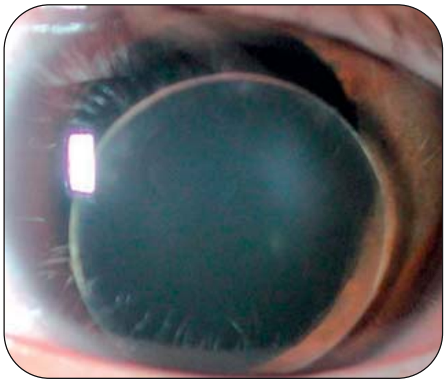

Fig. 6.2

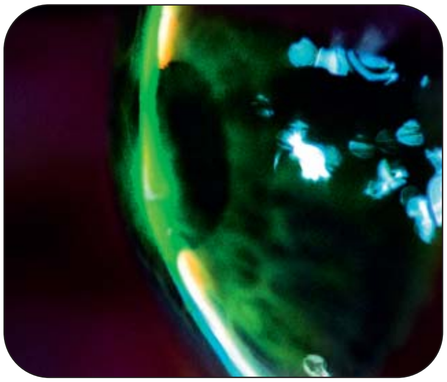

Fig. 6.3

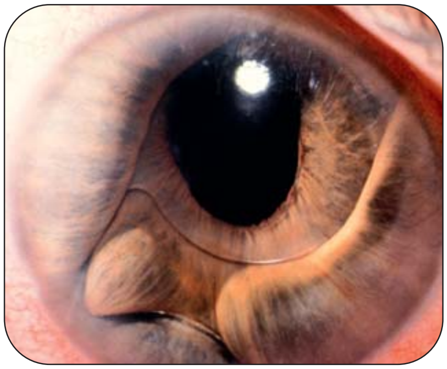

Fig. 6.4

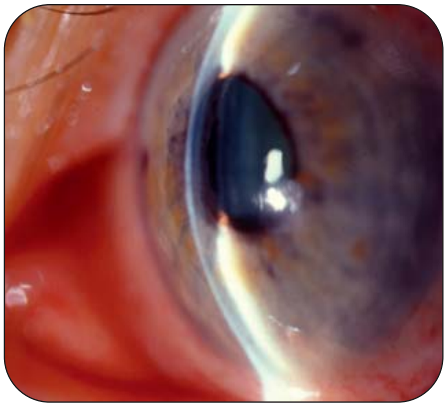

Fig. 6.5

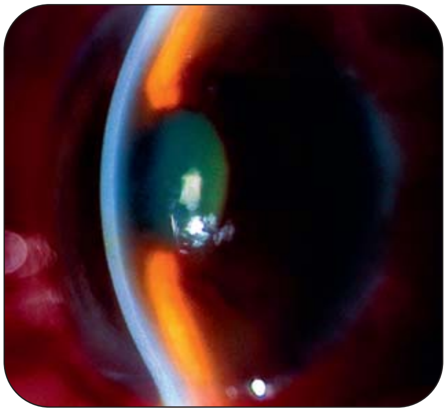

Fig. 6.6

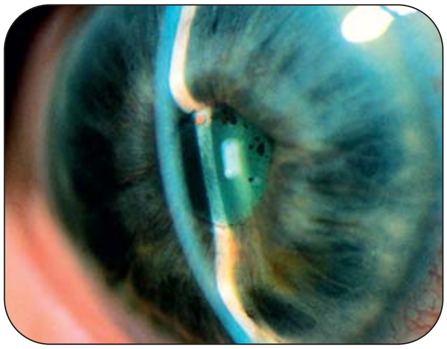

Fig. 6.7

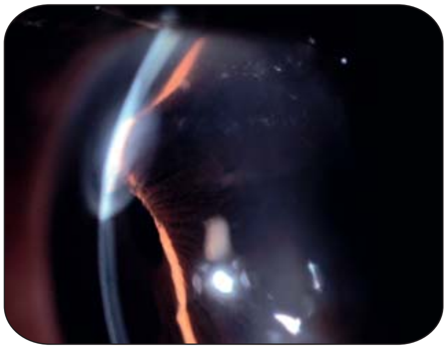

Fig. 6.8

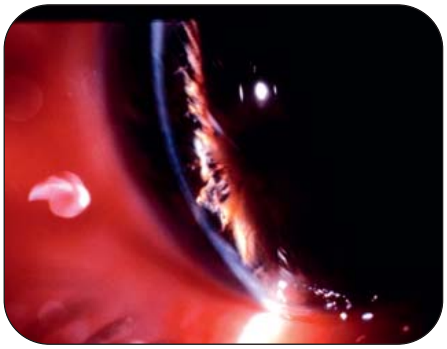

Fig. 6.9

Shallow chamber following filtration surgery

Leaking bleb

Signs

- *IOP* – low.
- *Bleb* – poor.
- *Iridectomy* – patent.
- *Seidel test* – positive (**Fig. 6.10**).

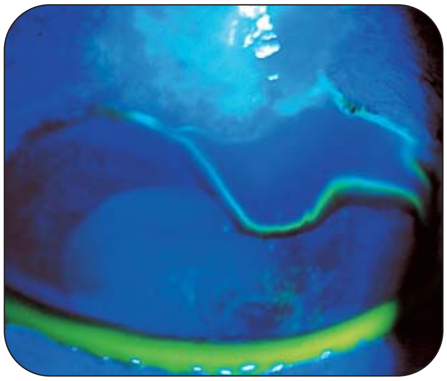

Fig. 6.10

Excessive filtration

Signs

- *IOP* – low.
- *Bleb* – large (**Fig. 6.11**).
- *Iridectomy* – patent.
- *Choroidal detachment* (**Fig. 6.12**).
- *Seidel test* – negative.

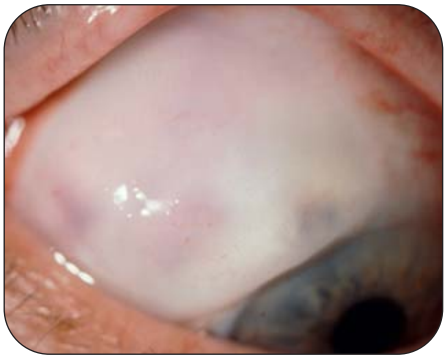
Fig. 6.11

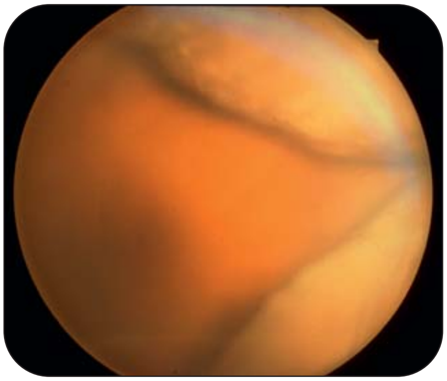
Fig. 6.12

Pupil block

Signs

- *IOP* – high.
- *Bleb* – flat.
- *Iridectomy* – non-patent.
- *Seidel test* – negative.

Malignant glaucoma

- *IOP* – very high.
- *Bleb* – absent (**Fig. 6.13**).
- *Iridectomy* – patent.
- *Seidel test* – negative.

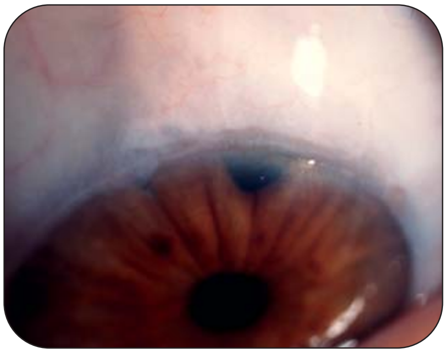
Fig. 6.13

INFLAMMATORY SIGNS

Anterior uveitis

Aqueous cells and flare

Signs

- See **Fig. 6.14**.

Cause

- Anterior uveitis.

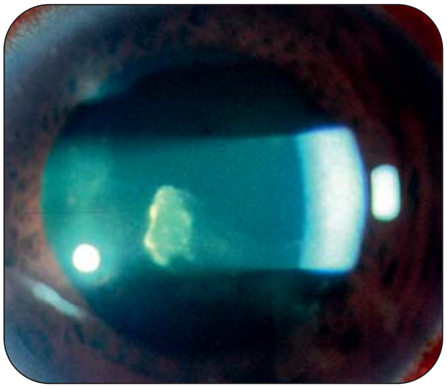
Fig. 6.14

Fibrinous exudate

Signs

- See **Fig. 6.15**.

Causes

- Severe non-infectious anterior uveitis, particularly in carriers of HLA-B27.
- Postoperative endophthalmitis.
- Sterile postoperative fibrinous reaction.

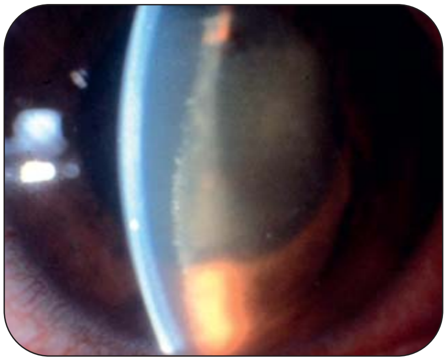

Fig. 6.15

Hypopyon

Signs

- See **Fig. 6.16**.

Causes

- Severe anterior uveitis – particularly associated with Behçet disease.
- Endophthalmitis.
- Severe infectious keratitis (**Fig. 6.17**).

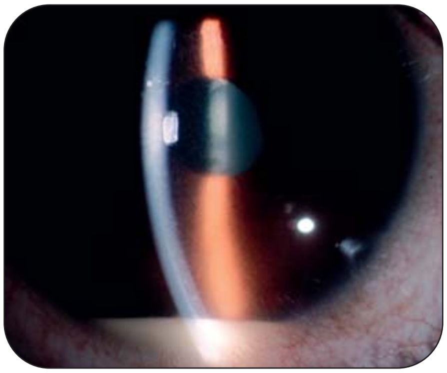

Fig. 6.16

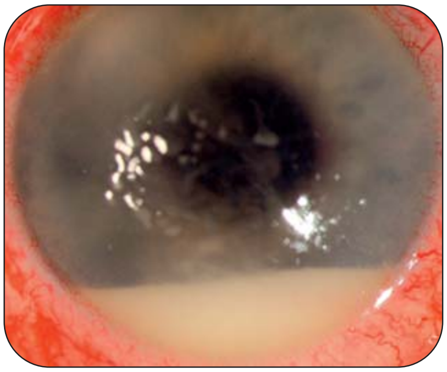

Fig. 6.17

Endophthalmitis

Acute postoperative

Presentation

- Usually during the first 5 days, most frequently following cataract surgery.

Signs

- Reduced visual acuity.
- Conjunctival injection and discharge.
- Corneal clouding.
- Fibrinous exudate and hypopyon (**Fig. 6.18**).
- Impaired red reflex.
- Vitreous cells and debris.

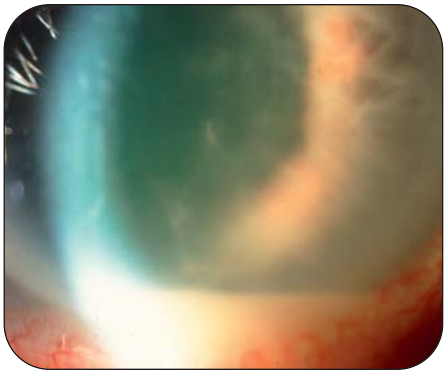

Fig. 6.18

Chronic postoperative

Presentation

- Several weeks or months after surgery. Caused by bacteria of low virulence.

Signs

- Mutton-fat keratic precipitates (**Fig. 6.19**).
- White fluffy plaque on posterior capsule (**Fig. 6.20**).

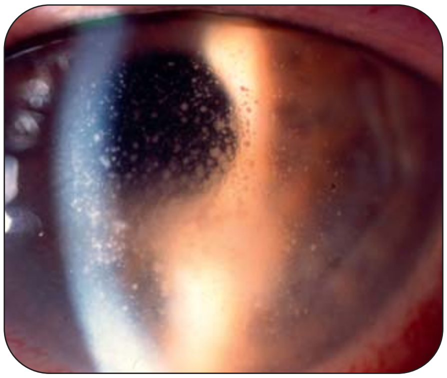

Fig. 6.19

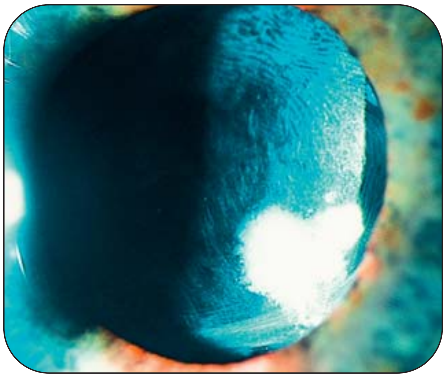

Fig. 6.20

Delayed-onset

Presentation

- Months or years following glaucoma filtration surgery. It is usually associated with a thin cystic filtering bleb and the use of adjunctive antimetabolites.

Signs

- White bleb surrounded by hyperaemia often associated with hypopyon (**Fig. 6.21**).

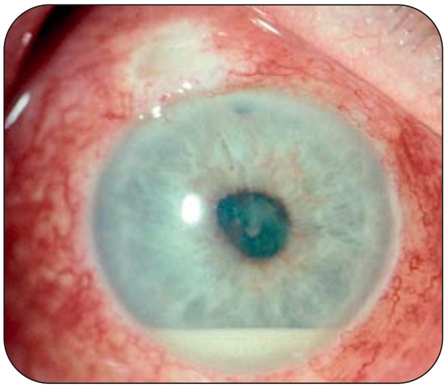

Fig. 6.21

Endogenous infectious

Causes

Immunocompromised status as in the following:

a. *Following splenectomy.*

b. *AIDS.*

c. *Intravenous drug abuse.*

d. *Diabetes mellitus.*

e. *Septic arthritis.*

NON-INFLAMMATORY CELLS

Hyphaema

Hyphaema describes the presence of red cells in the anterior chamber that may be caused by the following conditions.

Blunt ocular trauma

Trauma is a common cause of hyphaema due to bleeding from the iris or ciliary body.

Signs

- The red cells sediment inferiorly to form a 'fluid level' (**Fig. 6.22**).

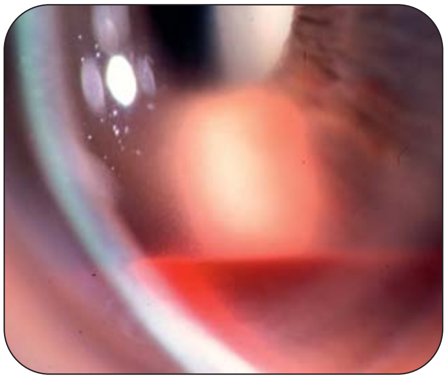

Fig. 6.22

Post-surgical

Mild hyphaema may occasionally occur following cataract or filtration surgery.

Causes of spontaneous hyphaema

a. *Herpes simplex and herpes zoster anterior uveitis.*

b. *Rubeosis iridis* (**Fig. 6.23**).

c. *Blood dyscrasias* associated with thrombocytopenia.

d. *Iris tumours*, particularly juvenile xanthogranuloma, and occasionally malignant primary and metastatic tumours (**Fig. 6.24**).

e. *Pupillary vascular (Cobb) tufts* (see Fig. 8.65).

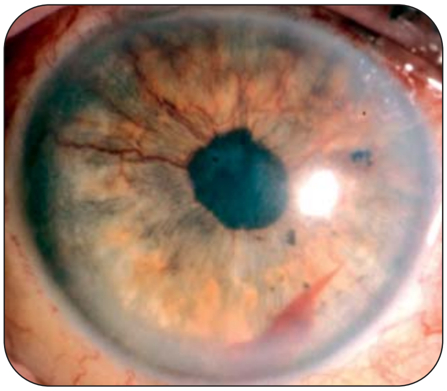

Fig. 6.23

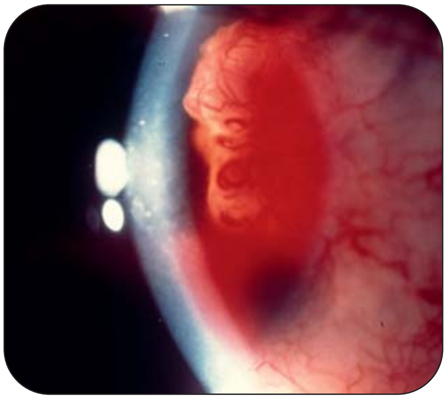

Fig. 6.24

Pseudo-hypopyon

Leukaemia cells

Signs

- Leukaemic cells may occasionally infiltrate the iris and anterior chamber (**Fig. 6.25**).

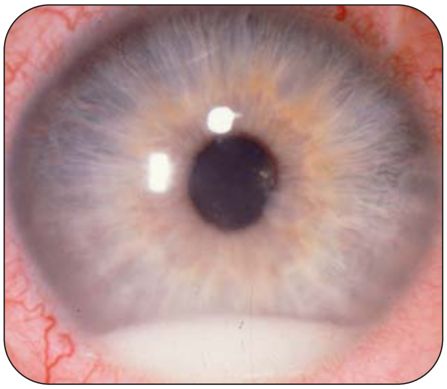

Fig. 6.25

Retinoblastoma

Signs

- Retinoblastoma may rarely invade the iris and anterior chamber (**Fig. 6.26**), particularly in older children.

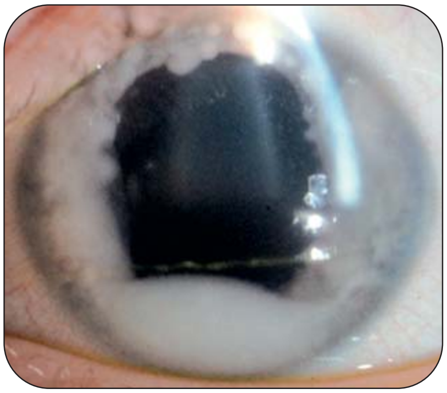

Fig. 6.26

Phacolytic glaucoma

Signs

- Phacolytic glaucoma occurs in eyes with hypermature cataracts in which white particles in the aqueous may form a pseudo-hypopyon (**Fig. 6.27**).

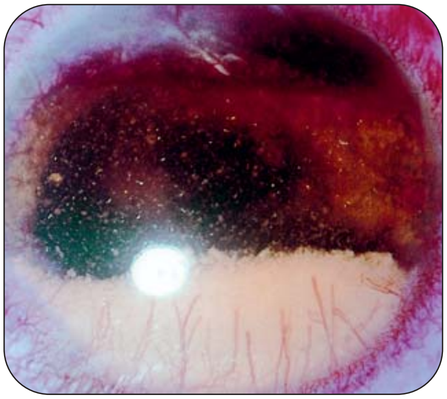

Fig. 6.27

Emulsified silicone oil

Signs

- Silicone oil may occasionally gain access to the anterior chamber and appear as an upside-down pseudo-hypopyon (**Fig. 6.28**).

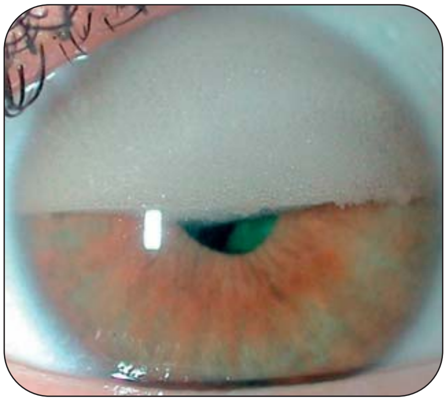

Fig. 6.28

Intraocular pressure and angle

7

TRABECULAR HYPERPIGMENTATION 153

Pseudoexfoliation syndrome 153
Pigment dispersion syndrome 153
Megalocornea 154
Naevus of Ota 154
Iris melanoma 155
Angle recession 155
Other causes 155

ANGLE CLOSURE 156

With pupil block 156
Primary 156
Inflammatory 156
Lens-induced 156

Without pupil block 157
Inflammatory 157
Rubeotic 157
Iridocorneal endothelial syndrome 157
Ciliary body tumour 158
Aniridia 158
Axenfeld–Rieger anomaly 158

ACUTE ELEVATION OF INTRAOCULAR PRESSURE 159

Primary angle closure 159
Phacomorphic glaucoma 159
Neovascular glaucoma 159
Glaucomatocyclitic crisis (Posner–Schlossman syndrome) 159
Secondary pupil block 160
Other causes 160

CHILDHOOD GLAUCOMA 160

Primary glaucoma 160
Isolated trabeculodysgenesis (primary congenital glaucoma) 160
Iridodysgenesis 161
Corneodysgenesis 161
Sturge–Weber syndrome 161
Other causes 161

Secondary glaucoma 162
Intraocular inflammation 162
Intraocular tumours 162
Ectopia lentis 162
Other causes 162

TRABECULAR HYPERPIGMENTATION

Pseudoexfoliation syndrome

Definition

- An uncommon, usually bilateral, condition that predisposes to glaucoma. It typically affects the elderly.

Signs

- Hyperpigmentation is most marked inferiorly. The pigment lies on the surface of the trabeculum and has a patchy distribution (**Fig. 7.1**).
- A scalloped band of pigment may extend anterior to Schwalbe line (Sampaolesi line).
- The trabeculum may also show 'dandruff-like' deposits of pseudoexfoliative material (PEM).

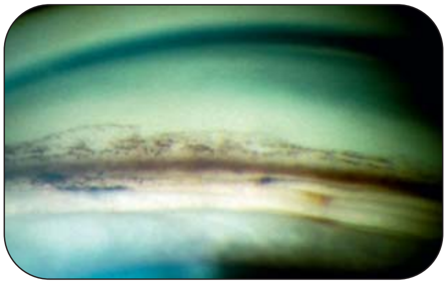

Fig. 7.1

Look for

- PEM on the pupil margin and sphincter atrophy giving rise to 'moth-eaten' and transillumination defects (see Fig. 8.79).
- PEM on the anterior lens surface (**Fig. 7.2**).
- Mild aqueous flare.
- Trabecular block glaucoma.

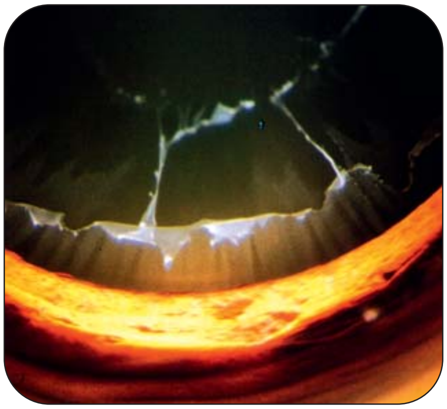

Fig. 7.2

Pigment dispersion syndrome

Definition

- An uncommon, bilateral, condition that predisposes to glaucoma. It typically affects young myopic males.

Signs

- Hyperpigmentation has a homogeneous appearance and forms a dense band involving the entire circumference of the meshwork uniformly (**Fig. 7.3**).
- The pigment is most marked over the posterior trabeculum. It is finer than in pseudoexfoliation and appears to lie on and within the trabecular meshwork.

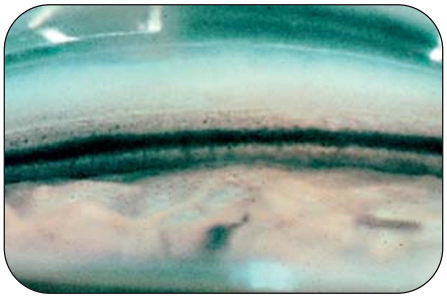

Fig. 7.3

Look for

- Fine pigment deposits on the endothelium (Krukenberg spindle – **Fig. 7.4**).
- Mid-peripheral radial slit-like transillumination defects (see Fig. 8.77).
- Partial loss of the pupillary ruff.
- Very deep anterior chamber.
- Pigment granules on the anterior iris surface.
- Trabecular block glaucoma.

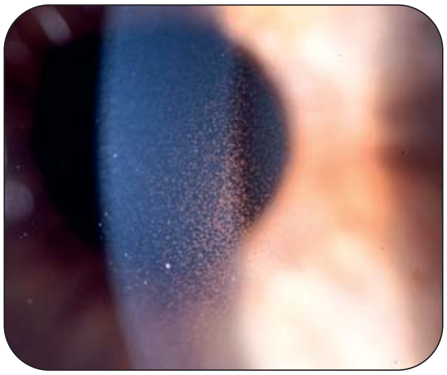

Fig. 7.4

Megalocornea

Definition

- A rare bilateral XL condition.

Signs

- Hyperpigmentation is similar to that in pigment dispersion syndrome and is associated with a very wide angle (**Fig. 7.5**).

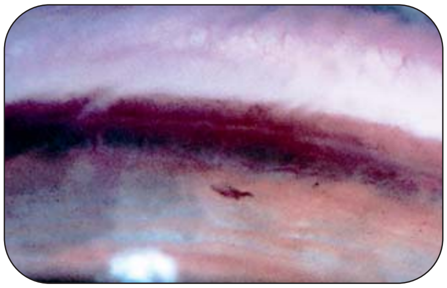

Fig. 7.5

Look for

- Enlarged but clear cornea that has a horizontal diameter ≥ 13 mm.
- Very deep anterior chamber (see Fig. 6.1).
- Glaucoma is uncommon.

Naevus of Ota

Definition

- A rare unilateral congenital condition.

Signs

- Hyperpigmentation homogeneously involves the entire circumference of the trabeculum (**Fig. 7.6**).

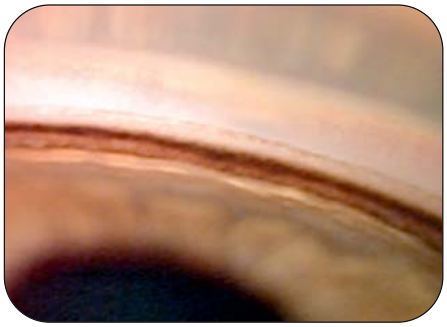

Fig. 7.6

Look for

- Hyperpigmentation of facial skin (see Fig. 1.47).
- Episcleral pigmentation (see Fig. 3.66).
- Heterochromia iridis (see Fig. 8.97).
- Iris mammillations (see Fig. 8.42).
- Fundus hyperpigmentation (**Fig. 7.7**).
- Trabecular block glaucoma.

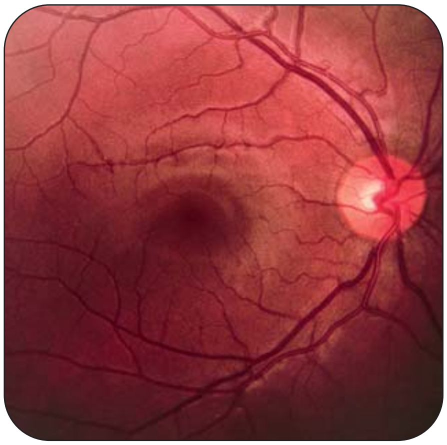

Fig. 7.7

Iris melanoma

Melanoma may cause trabecular hyperpigmentation by two mechanisms.

a. *Direct infiltration* of the meshwork by neoplastic cells is by far the most common (**Fig. 7.8**).

b. *Indirect involvement* by macrophages that have ingested pigment and tumour cells, similar to pigment dispersion syndrome (**Fig. 7.9**).

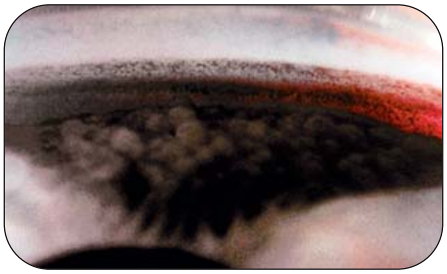

Fig. 7.8

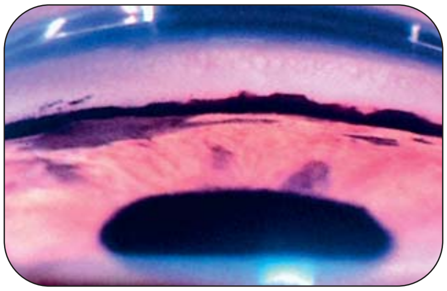

Fig. 7.9

Angle recession

Cause

- Severe blunt ocular trauma.

Signs

- Localised trabecular hyperpigmentation and irregular widening of the ciliary body band (**Fig. 7.10**).

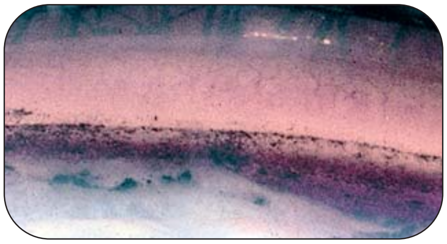

Fig. 7.10

Look for

- Rupture of the iris sphincter (**Fig. 7.11**).
- Iridodialysis.
- Lens dislocation.
- Cataract.
- Late glaucoma.

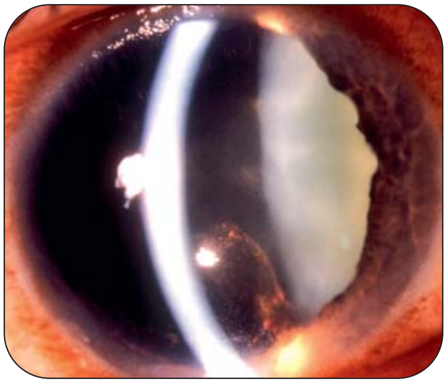

Fig. 7.11

Other causes

a. *Pseudophakic pigment dispersion* occurs as the result of rubbing of the haptics and optics of a posterior chamber IOL against the iris epithelium (see Fig. 8.78).

b. *Subacute angle closure* may be associated with heavy trabecular pigmentation where the iris root has been in contact with the angle.

c. *Anterior uveitis* may give rise to scattered trabecular pigmentation.

ANGLE CLOSURE

With pupil block

Primary

Pathogenesis

- Angle closure occurs in anatomically predisposed eyes.
- Aqueous flow from the posterior to the anterior chamber is obstructed by relative pupil block resulting in iris bombé and closure of the angle by the peripheral iris (**Fig. 7.12**).

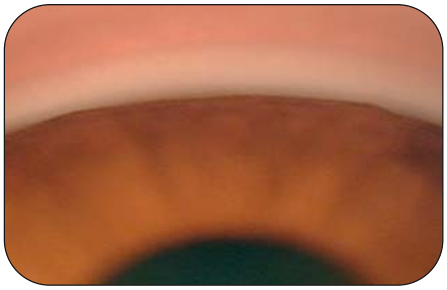

Fig. 7.12

Inflammatory

Pathogenesis

- Angle closure occurs in eyes with chronic anterior uveitis in which seclusio pupillae obstructs aqueous flow from the posterior to the anterior chamber.
- Increased pressure in the posterior chamber produces iris bombé and apposition of the peripheral iris to the trabeculum and peripheral cornea (**Fig. 7.13**).

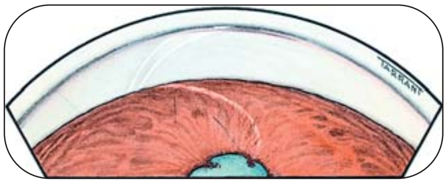

Fig. 7.13

Lens-induced

Lens-induced angle closure occurs in the following circumstances:

a. *Phacomorphic glaucoma* caused by an intumescent (swollen) lens (see Fig. 7.25).

b. *Total dislocation* of the lens into the anterior chamber either caused by blunt ocular trauma or spontaneous dislocation of a microspherical lens (see Fig. 6.2).

c. *Incarceration* of a microspherical lens in the pupil in which only part of the zonule has been disrupted so that the intact zonule acts like a hinge (**Fig. 7.14**).

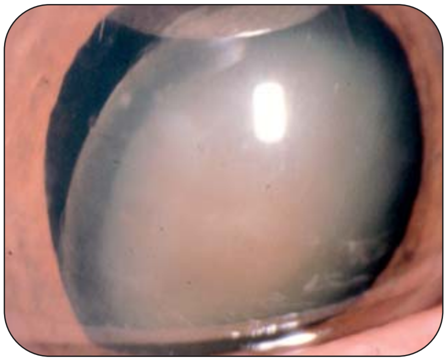

Fig. 7.14

Without pupil block

Inflammatory

Pathogenesis

- Angle closure is caused by organisation and contraction of inflammatory cells and debris in the angle causes gradual and progressive formation of peripheral anterior synechiae (PAS – **Fig. 7.15**).

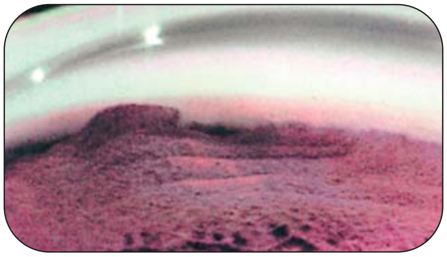

Fig. 7.15

Rubeotic

Pathogenesis

- Angle closure is caused by contraction of fibrovascular tissue and circumferential pulling of the peripheral iris over the trabeculum in a zipper-like manner with the formation of PAS (**Fig. 7.16**).

Look for

- Ischaemic central retinal vein occlusion.

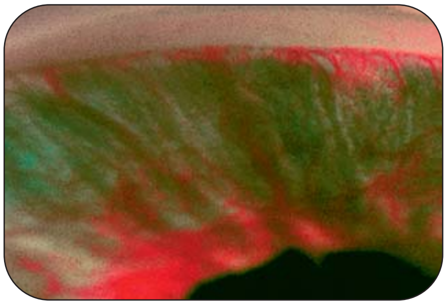

Fig. 7.16

Iridocorneal endothelial syndrome

Pathogenesis

- Angle closure is caused by the gradual formation of broad PAS that may extend anterior to Schwalbe line (**Fig. 7.17**).

Look for

- Iris atrophy, corectopia and pseudopolycoria (**Fig. 7.18**).
- Diffuse iris naevus and multiple nodules (see Fig. 8.43).
- Hammered-silver endothelial changes.
- Glaucoma.

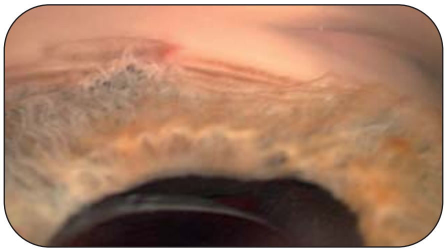

Fig. 7.17

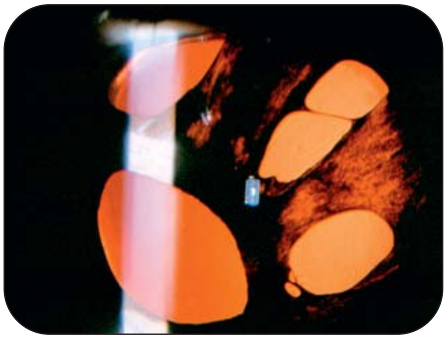

Fig. 7.18

Ciliary body tumour

Pathogenesis

- Angle closure is caused by anterior displacement of the iris (**Fig. 7.19**).

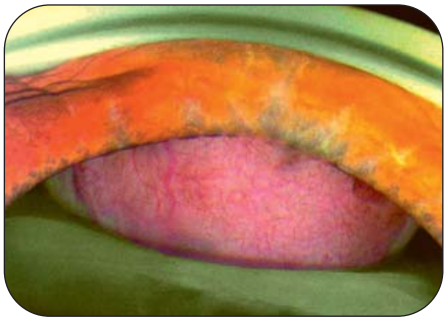

Fig. 7.19

Aniridia

Pathogenesis

- Angle closure is caused by pulling forward of rudimentary iris tissue over the trabeculum (**Fig. 7.20**).

Look for

- Tear film instability and dry eye.
- Conjunctivalisation of peripheral cornea.
- Lens subluxation (**Fig. 7.21**).
- Foveal hypoplasia.
- Glaucoma.

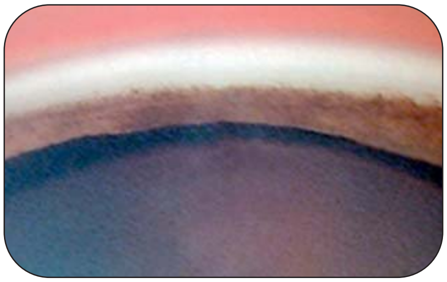

Fig. 7.20

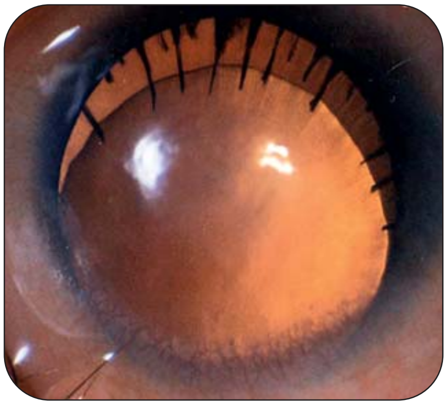

Fig. 7.21

Axenfeld–Rieger anomaly

Pathogenesis

- Angle closure is caused by PAS consisting of broad sheets of iris stromal adherent to the cornea anterior to Schwalbe line (**Fig. 7.22**).

Look for

- Posterior embryotoxon (see Fig. 5.116).
- Iris shows ectropion uveae, diffuse stromal atrophy and corectopia (**Fig. 7.23**).
- Glaucoma is common.

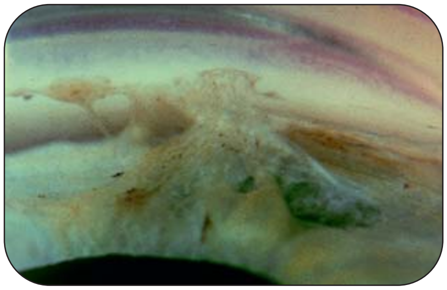

Fig. 7.22

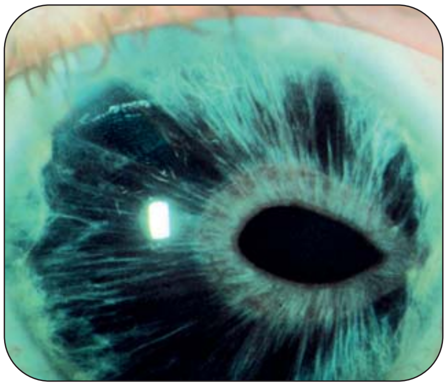

Fig. 7.23

ACUTE ELEVATION OF INTRAOCULAR PRESSURE

Primary angle closure

Presentation

- Abrupt onset of unilateral visual loss and pain in an elderly person.

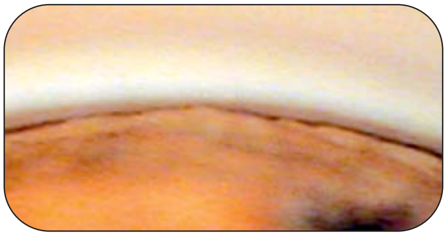
Fig. 7.24

Signs

- Corneal epithelial oedema.
- Closed angle and shallow anterior chamber.
- Fellow eye shows a narrow angle (**Fig. 7.24**) and a shallow anterior chamber.

Phacomorphic glaucoma

Pathogenesis

- A unilateral condition caused by an intumescent (swollen) lens. Patients have a long history of visual loss before the acute event.

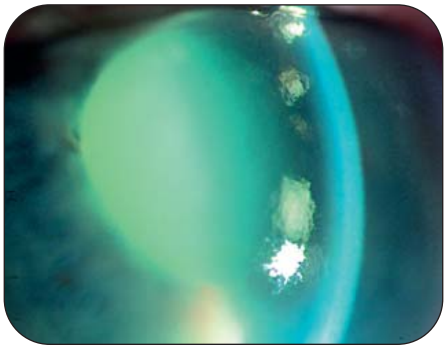
Fig. 7.25

Signs

- Corneal oedema, angle closure and advanced cataract (**Fig. 7.25**).
- Fellow eye shows normal angle and anterior chamber depth.

Neovascular glaucoma

Pathogenesis

- A unilateral condition caused by rubeosis iridis, most frequently secondary to ischaemic central retinal vein occlusion. Patients have a long history of visual loss before the acute event.

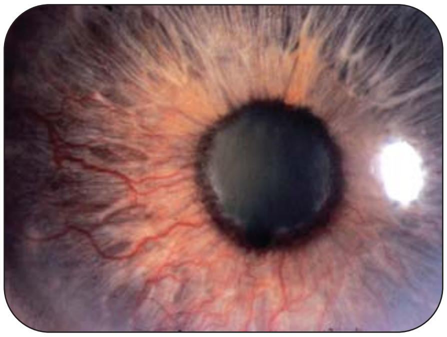
Fig. 7.26

Signs

- Advanced rubeosis iridis (**Fig. 7.26**).
- Fellow eye shows a normal angle and anterior chamber depth.

Glaucomatocyclitic crisis (Posner–Schlossman syndrome)

Description

- A rare unilateral condition characterised by severe recurrent elevation of intraocular pressure.

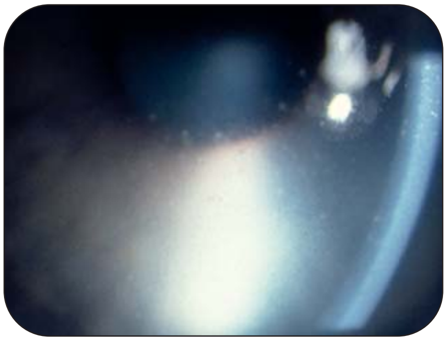
Fig. 7.27

Signs

- Small keratic precipitates (**Fig. 7.27**).
- Both eyes show normal angles and anterior chamber depths.

Secondary pupil block

Pathogenesis

- Seclusio pupillae caused by chronic anterior uveitis.

Signs

- Closed angle and iris bombé (see Fig. 7.13).
- Fellow eye shows a normal angle (**Fig. 7.28**) and anterior chamber depth.

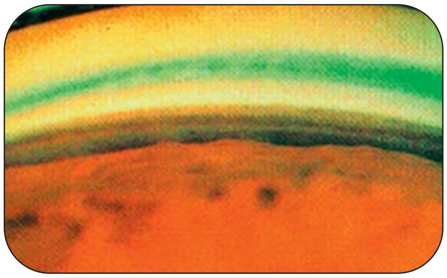

Fig. 7.28

Other causes

a. *Pigment dispersion syndrome* may occasionally cause sudden elevation of IOP and blurred vision, particularly after strenuous physical exertion.

b. *Pseudoexfoliation syndrome* is occasionally associated with unilateral acute pressure rise despite an open angle.

CHILDHOOD GLAUCOMA

Primary glaucoma

Isolated trabeculodysgenesis (primary congenital glaucoma)

Presentation

- Within the first three years of life. It is bilateral in 75% of cases but involvement is frequently asymmetric.

Signs

- Photophobia and blepharospasm (**Fig. 7.29**).
- Corneal haze and buphthalmos (see Fig. 5.7).
- The trabeculum and the ciliary body band are obscured by translucent amorphous material (**Fig. 7.30**).

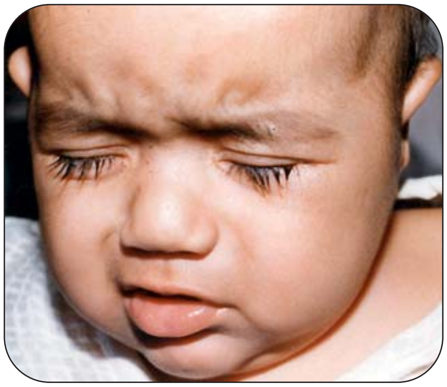

Fig. 7.29

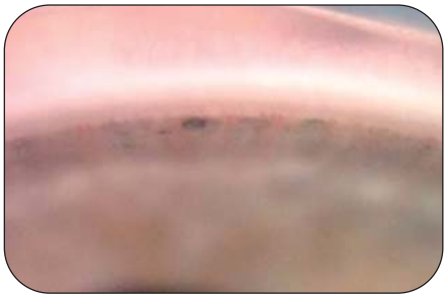

Fig. 7.30

Iridodysgenesis

Associations

- Aniridia, which is frequently associated with severe glaucoma.
- Congenital ectropion uveae (**Fig. 7.31**).

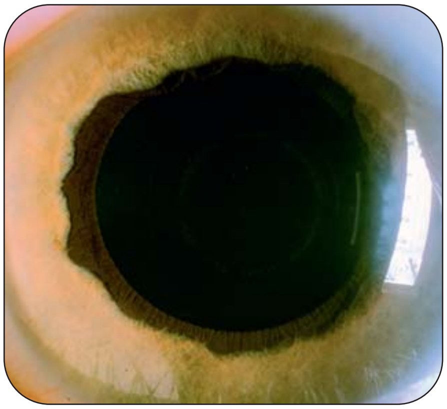

Fig. 7.31

Corneodysgenesis

Associations

- Axenfeld–Rieger anomaly and syndrome (see Figs 7.22 and 7.23).
- Peters anomaly (see Fig. 6.8).
- Microcornea (see Fig. 5.1).

Sturge–Weber syndrome

- Glaucoma occurs in about 30% of patients ipsilateral to the facial haemangioma which is occasionally bilateral (**Fig. 7.32**).
- The pressure rise is caused either by an isolated trabeculodysgenesis or raised episcleral pressure associated with an arteriovenous communication in an episcleral haemangioma.

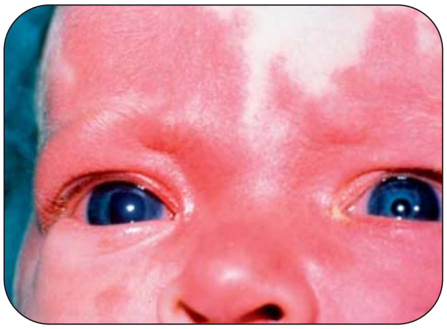

Fig. 7.32

Other causes

a. *NF1* is associated with glaucoma in about 50% of patients with a plexiform neurofibroma of the upper eyelid (**Fig. 7.33**) or those with facial hemiatrophy.

b. *Oculocerebrorenal (Lowe) syndrome* is a very rare condition in which all children have cataract and 50% glaucoma.

c. *Mucopolysaccharidoses* such as Hurler syndrome are occasionally associated with glaucoma.

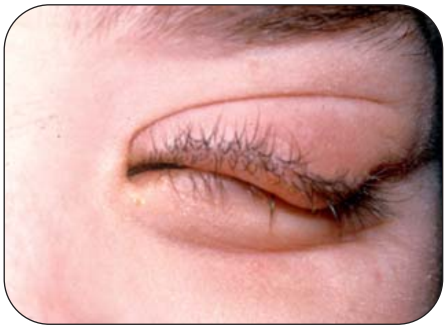

Fig. 7.33

Secondary glaucoma

Intraocular inflammation

- Chronic anterior uveitis most frequently associated with juvenile idiopathic arthritis.
- Congenital rubella may cause glaucoma as well as keratitis, cataract, iris atrophy and retinopathy.

Intraocular tumours

- Retinoblastoma may cause buphthalmos (**Fig. 7.34**).
- Juvenile xanthogranuloma which may cause spontaneous hyphaema that may result in elevation of intraocular pressure.

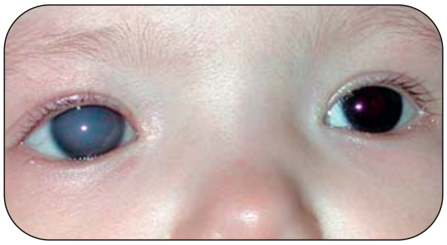

Fig. 7.34

Ectopia lentis

Lens subluxation associated with the following conditions may cause elevation of intraocular pressure.

a. *Blunt trauma.*

b. *Marfan syndrome.*

c. *Weill–Marchesani syndrome.*

d. *Stickler syndrome.*

e. *Ehlers–Danlos syndrome.*

f. *Homocystinuria.*

g. *Sulfite oxidase deficiency.*

h. *Hyperlysinaemia.*

i. *Aniridia.*

Other causes

a. *Aphakia* following surgery for congenital cataract is now a rare condition due to refined surgical techniques.

b. *Persistent anterior fetal vasculature* (see Fig. 9.19) is a unilateral condition that may be associated with swelling of the lens and secondary angle closure.

Iris 8

ANTERIOR UVEITIS 165

Clinical features 165

Acute 165

Chronic 165

Anterior uveitis without systemic associations 166

Fuchs syndrome 166

HLA-B27 associated 167

Lens-induced 167

Drug-induced 167

Posner–Schlossman syndrome 167

Systemic associations of anterior uveitis 167

Ankylosing spondylitis 167

Reiter syndrome 168

Psoriatic arthritis 169

Behçet disease 169

Acute sarcoidosis 170

Chronic sarcoidosis 170

Vogt–Koyanagi–Harada syndrome 170

Juvenile idiopathic arthritis 171

Inflammatory bowel disease 171

Relapsing polychondritis 171

Other associations 172

IRIS SPOTS AND NODULES 172

Solitary 172

Naevus 172

Melanoma 172

Leiomyoma 173

Melanocytoma 173

Metastatic carcinoma 173

Forward extension of ciliary body melanoma 174

Multiple 174

Freckles 174

Brushfield spots 174

Lisch nodules 174

Mammillations 175

Iris naevi in Cogan–Reese syndrome 175

Inflammatory nodules 175

Tapioca melanoma 176

Multiple iris metastases 176

Retinoblastoma invading the iris 176

Iris flocculi 177

CYSTIC LESIONS 177

Primary 177

Epithelial 177

Stromal 177

Secondary 178

Implantation 178

Miotic 178

Parasitic 178

ECTROPION UVEAE 179

Congenital 179

Acquired 179

Associated with rubeosis iridis 179

Associated with iris tumours 179

RUBEOSIS IRIDIS 180

IRIS ATROPHY AND TRANSLUCENCY 181

Fuchs syndrome 181

Albinism 181

Axenfeld–Rieger anomaly 182

Post acute angle-closure 183

Iridoschisis 183

Pigment dispersion syndrome 183

Pseudophakic pigment dispersion 184

Pseudoexfoliation syndrome 184

Diabetic iridopathy 184

Herpes zoster and simplex anterior uveitis 185

Lepromatous iritis 185

Rubella iritis 185

FULL-THICKNESS DEFECT OR ABSENCE 186

Aniridia 186

Iris coloboma 187

Axenfeld–Rieger anomaly 187

Iridocorneal-endothelial syndrome 187

Blunt trauma 188

Laser iridotomy 188

HETEROCHROMIA IRIDIS 189

Congenital 189

Isolated simple 189

Waardenburg syndrome 189

Parry–Romberg syndrome 189

Horner syndrome 190

Oculodermal melanocytosis (naevus of Ota) 190

Sturge–Weber syndrome 190

Hirschsprung disease 190

Acquired 191

Fuchs syndrome 191

Unilateral use of a prostaglandin analogue 191

Ocular siderosis 191

Diffuse pigmented iris tumours 191

ANTERIOR UVEITIS

Clinical features

Acute

Signs

- *Injection* – intense and circumcorneal (ciliary – **Fig. 8.1**).
- *Pupil* – small.
- *Keratic precipitates* (KP) – endothelium dusting by myriads of cells.
- *Flare and cells* – often intense (**Fig. 8.2**).
- *Fibrinous exudate* – if severe (**Fig. 8.3**).
- *Hypopyon* – if very severe.
- *Iris* – usually unremarkable; occasionally shows dilated capillaries (see Fig. 8.63).

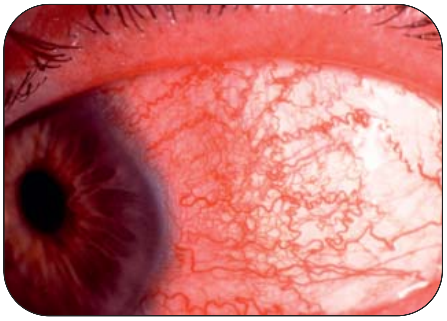
Fig. 8.1

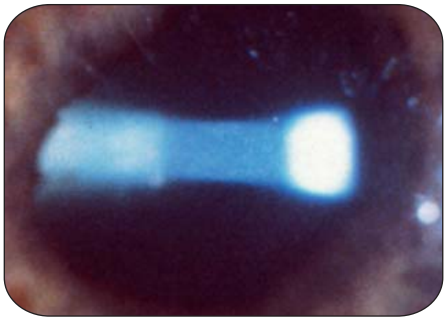
Fig. 8.2

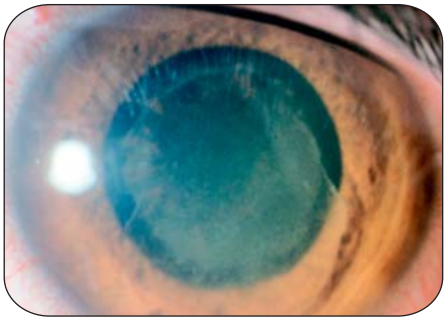
Fig. 8.3

Complications

- *Posterior synechiae (PS)* – rare at presentation but may form later.
- *Cataract* – absent.
- *Glaucoma* – rare.

Chronic

Signs

- *Injection* – mild or absent.
- *Pupil* – unremarkable.
- *KP* – mutton-fat in granulomatous disease (**Fig. 8.4**).
- *Flare and cells* – variable.
- *Fibrinous exudate* – absent.
- *Hypopyon* – rare.
- *Iris* – nodules in granulomatous disease, especially sarcoidosis (**Fig. 8.5**).

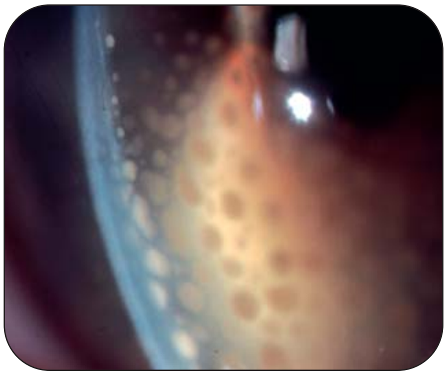
Fig. 8.4

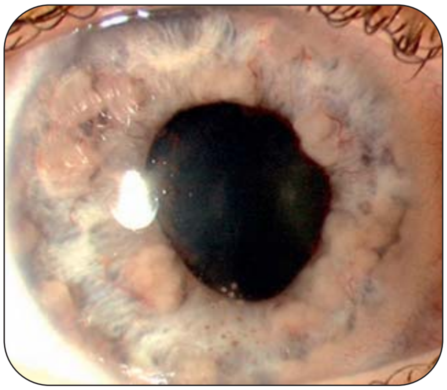
Fig. 8.5

Complications

- *PS* – common at presentation (**Fig. 8.6**).
- *Cataract* – rare at presentation but may develop later.
- *Glaucoma* – rare at presentation but may develop later.

Table 8.1 **Causes of granulomatous uveitis**

1. **Systemic non-infectious causes**
   - Chronic sarcoidosis
   - Vogt–Koyanagi–Harada syndrome
   - Multiple sclerosis
2. **Systemic infections**
   - Toxoplasmosis
   - Lyme disease
   - Tuberculosis
   - Syphilis
3. **Local ocular causes**
   - Propionibacterium acnes endophthalmitis
   - Sympathetic ophthalmitis
   - Phacoanaphylactic uveitis

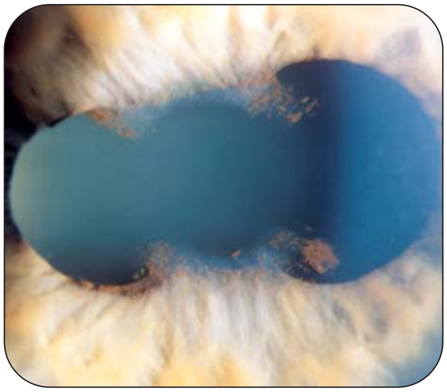

Fig. 8.6

Anterior uveitis without systemic associations

Fuchs syndrome

Definition

- A unilateral idiopathic chronic anterior uveitis (CAU) with an insidious onset that typically presents in early adult life.

Signs

- *Injection* – absent.
- *Pupil* – unremarkable.
- *KP* – small, grey-white, scattered throughout endothelium and frequently associated with feathery fibrin filaments (**Fig. 8.7**).
- *Flare and cells* – mild to moderate.
- *Fibrinous exudate* – absent.
- *Hypopyon* – absent.
- *Iris* – small stromal nodules (**Fig. 8.8**), small pupillary nodules and stromal atrophy (**Fig. 8.9**), and heterochromia.
- *Vitritis* – may be severe.
- *Angle* – fine twig-like vessels over the trabeculum (**Fig. 8.10**).

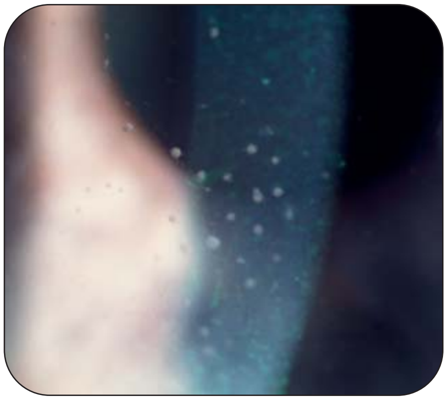

Fig. 8.7

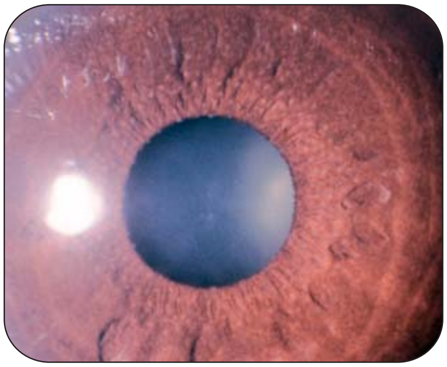

Fig. 8.8

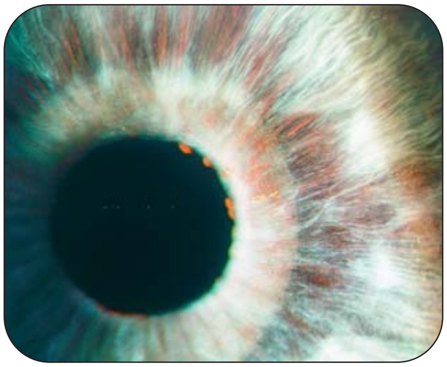

Fig. 8.9

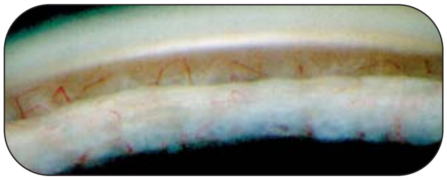

Fig. 8.10

Complications

- *PS* – absent.
- *Cataract* – common at presentation.
- *Glaucoma* – rare at presentation but common in long-standing cases.

HLA-B27 associated

HLA-B27 patients may suffer from severe recurrent acute anterior uveitis (AAU) often associated with a fibrinous exudate (see Fig. 8.3).

Lens-induced

- Phacoanaphylactic endophthalmitis – bilateral CAU.
- Phacogenic – unilateral CAU.

Drug-induced

Drugs that may cause bilateral AAU are:

- Rifabutin.
- Cidofovir.

Posner–Schlossman syndrome

Definition

- A rare condition characterised by recurrent unilateral attacks of severe elevation of intraocular pressure associated with mild AAU and small KP (see Fig. 7.27).

Systemic associations of anterior uveitis

Ankylosing spondylitis

Incidence

- Severe recurrent AAU affects about 30% of cases and is frequently associated with a fibrinous exudate.

Look for

- HLA-B27 – positive in 90% of cases.
- Pain and stiffness in the lower back or buttocks, more marked in the mornings due to sacroiliitis.
- Rigid neck with lack of extension (**Fig. 8.11**).

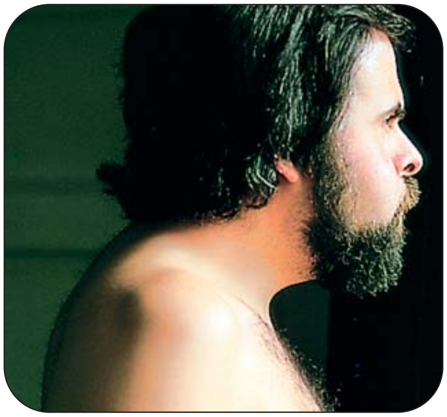

Fig. 8.11

- Limitation of spinal movements due to syndesmophyte formation and ossification of anterior spinal ligaments (bamboo spine – **Fig. 8.12**).

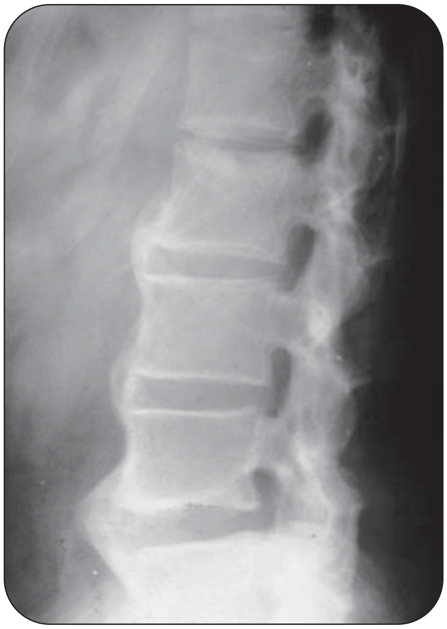

Fig. 8.12

Reiter syndrome

Incidence

- AAU is common but acute conjunctivitis is universal.

Look for

- HLA-B27 – positive in 85%.
- Urethritis is the presenting feature.
- Painless mouth ulceration.
- Asymmetrical arthritis most frequently involving the knees and ankles.
- Circinate balanitis (**Fig. 8.13**).
- Keratoderma blenorrhagica involving the palms and soles (see Fig. 3.40).
- Sacroiliitis, particularly in carriers of HLA-B27, resulting in irregularity and loss of cortical margins with subsequent sclerosis, narrowing and fusion (**Fig. 8.14**).

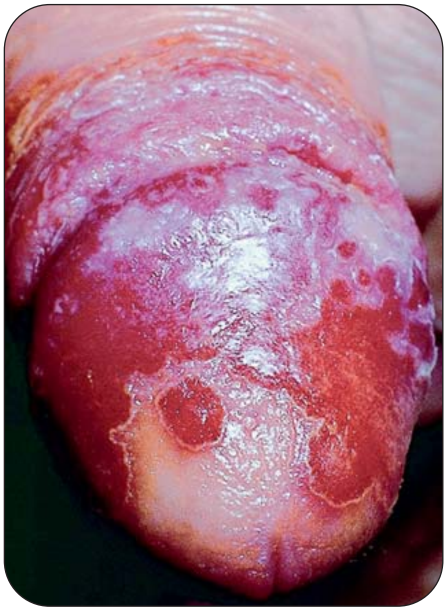

Fig. 8.13

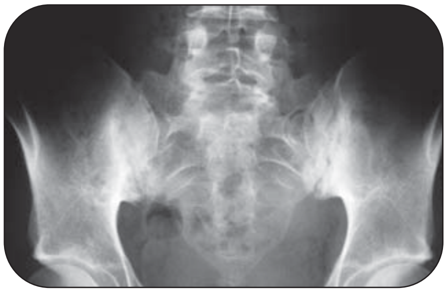

Fig. 8.14

Psoriatic arthritis

Incidence

- AAU is uncommon.

Look for

- HLA-B27 – uncommon.
- Well-demarcated, scaly patches on extensor surfaces.
- Asymmetric arthritis involving the knuckles and distal interphalangeal joints that may result in severe deformity (arthritis mutilans – **Fig. 8.15**).
- Nail dystrophy (**Fig. 8.16**).

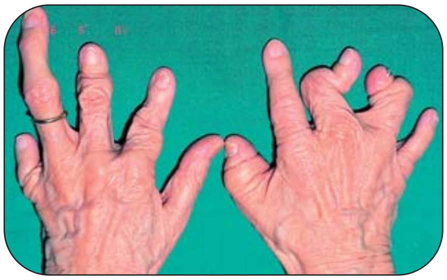

Fig. 8.15

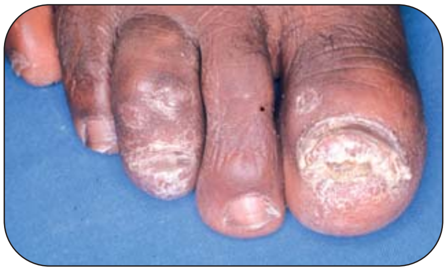

Fig. 8.16

Behçet disease

Incidence

- AAU associated with hypopyon is common but the eye usually lacks ciliary injection (**Fig. 8.17**).

Look for

- HLA-B51 – common.
- Recurrent painful oral (**Fig. 8.18**) and genital ulceration (see Figs 15.99 and 15.100).
- Cutaneous hypersensitivity (dermatographia – **Fig. 8.19**), erythema nodosum and acneiform lesions.
- Recurrent thrombophlebitis.

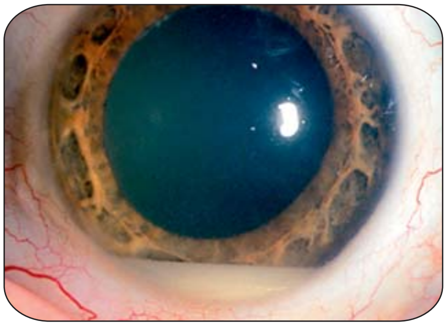

Fig. 8.17

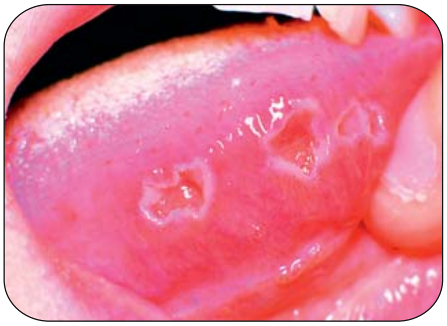

Fig. 8.18

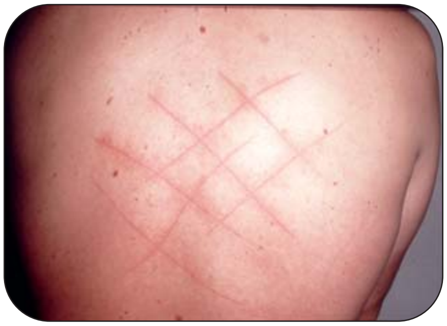

Fig. 8.19

Acute sarcoidosis

Incidence

- AAU is uncommon.

Look for

- Löfgren syndrome – fever, erythema nodosum (**Fig. 8.20**) and hilar lymphadenopathy (**Fig. 8.21**).
- Heerfordt syndrome – fever and parotid enlargement (**Fig. 8.22**).

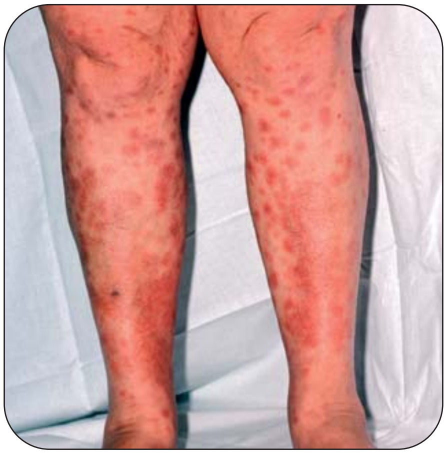

Fig. 8.20

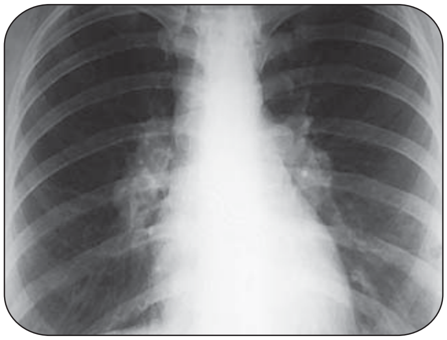

Fig. 8.21

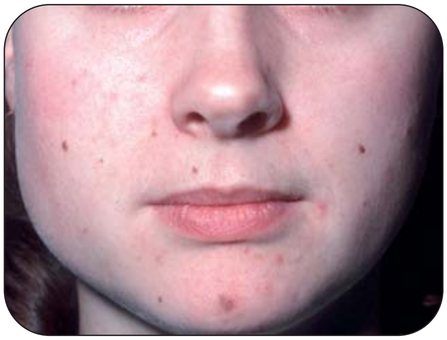

Fig. 8.22

Chronic sarcoidosis

Incidence

- Granulomatous anterior uveitis (GAU) is common.

Look for

- Cutaneous lesions – lupus pernio and granuloma (**Fig. 8.23**).
- Pulmonary disease – parenchymal infiltrates and fibrosis (**Fig. 8.24**).
- Neurological lesions – multifocal peripheral neuropathy with a predilection for the facial nerve, meningeal infiltration and CNS granulomas.

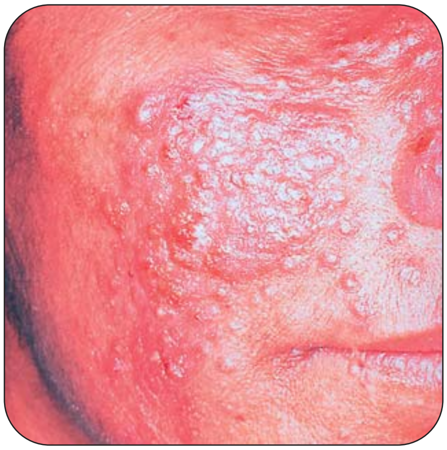

Fig. 8.23

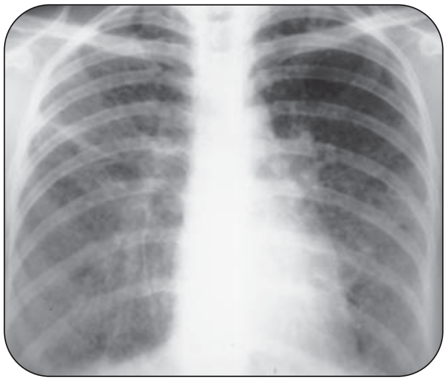

Fig. 8.24

Vogt–Koyanagi–Harada syndrome

Incidence

- Anterior uveitis which is initially acute and then becomes chronic and granulomatous is common.

Look for

- Hispanic, Japanese or pigmented individual.
- Alopecia, poliosis and vitiligo (**Fig. 8.25**).

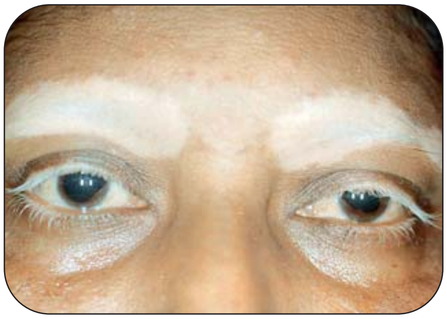

Fig. 8.25

Juvenile idiopathic arthritis

Incidence

- CAU is common in pauciarticular-onset disease and may lead to band keratopathy and cataract (**Fig. 8.26**).

Look for

- Child under the age of 16 years.
- Pauciarticular-onset arthritis most frequently involving the knees (**Fig. 8.27**).

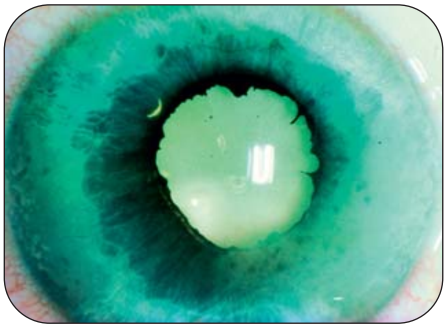
Fig. 8.26

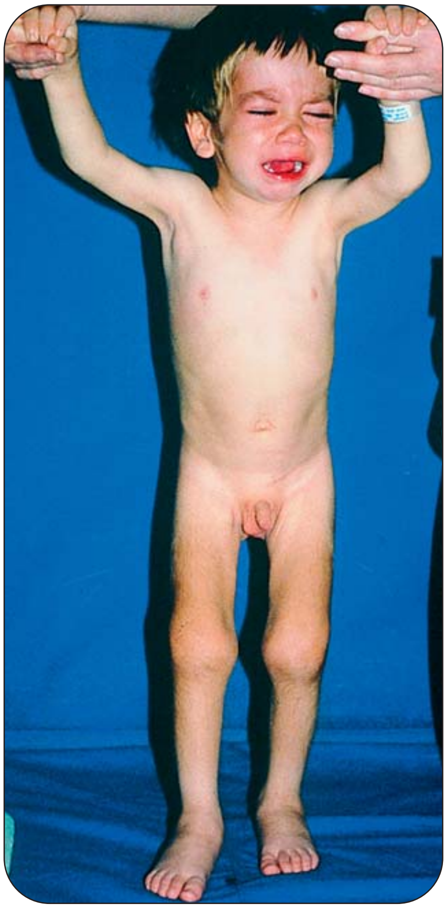
Fig. 8.27

Inflammatory bowel disease

Incidence

- Ulcerative colitis and Crohn disease may be associated with AAU particularly in patients with associated sarcoiliitis.

Look for

- Weight loss, fever and diarrhoea.
- Pyoderma gangrenosum (**Fig. 8.28**) and erythema nodosum.
- Anal tags and fistulae in Crohn disease.

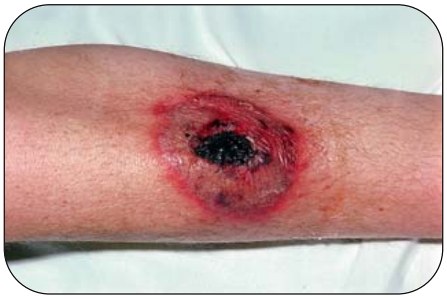
Fig. 8.28

Relapsing polychondritis

Incidence

- AAU is uncommon.

Look for

- Recurrent inflammation of pinnae.
- Collapse of nasal cartilage (**Fig. 8.29**).

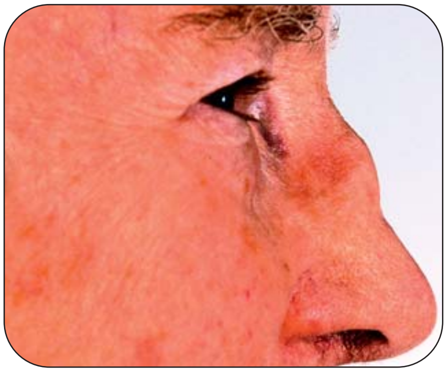
Fig. 8.29

Other associations

a. *Tuberculosis* – CAU and GAU.

b. *Acquired syphilis* – AAU and GAU.

c. *Tubulointerstitial nephritis* – CAU.

d. *IgA glomerulonephritis* – AAU.

e. *Leprosy* – AAU and CAU.

f. *Lyme disease* – GAU.

g. *Brucellosis* – CAU.

h. *Neonatal-onset multisystem inflammatory disease* – CAU.

IRIS SPOTS AND NODULES

Solitary

Naevus

Signs

- Unilateral flat or slightly elevated pigmented lesion involving the superficial layers of the iris (**Fig. 8.30**).
- Mild pupillary distortion and ectropion uveae are occasionally seen.
- In the Cogan–Reese syndrome the naevus is more diffuse, obscures the normal pattern of iris crypts and gives rise to hyperchromic heterochromia (**Fig. 8.31**).

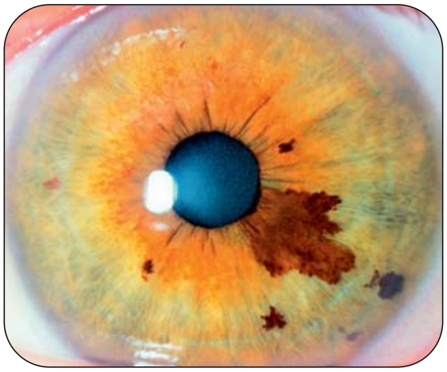

Fig. 8.30

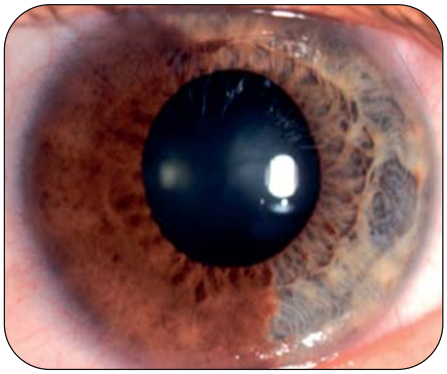

Fig. 8.31

Melanoma

Definition

- A very slow-growing tumour of relatively low malignancy which typically presents during the 5th decade of life.

Signs

- Pigmented or non-pigmented nodule, at least 3 mm in diameter and 1 mm in thickness, located in the inferior half of the iris (**Fig. 8.32**).

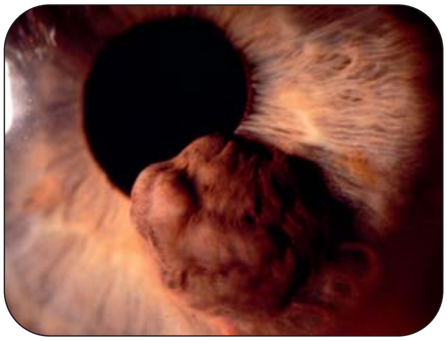

Fig. 8.32

- Vascularisation is easier to detect in a non-pigmented tumour (**Fig. 8.33**).
- Pupillary distortion, ectropion uveae and secondary lens opacities.
- Angle involvement may give rise to raised intraocular pressure (see Figs 7.8 and 7.9).

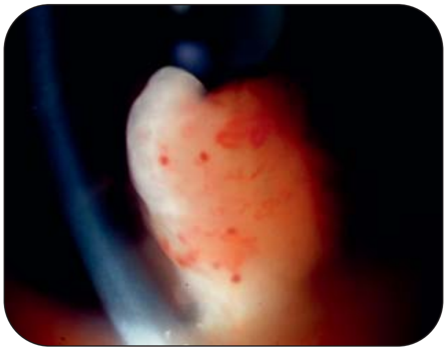

Fig. 8.33

Leiomyoma

Signs

- Similar to an amelanotic melanoma except that it is not confined to the inferior part of the iris (**Fig. 8.34**).

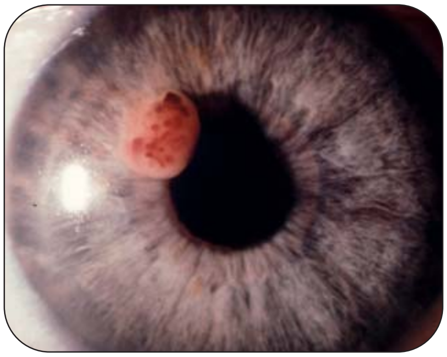

Fig. 8.34

Melanocytoma

Signs

- Black friable nodule with a mossy granular surface that has a predilection for the inferior iris root (**Fig. 8.35**).
- Aqueous seeding may cause elevation of intraocular pressure.

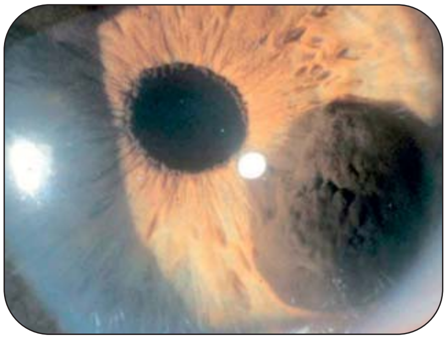

Fig. 8.35

Metastatic carcinoma

Signs

- Fast-growing amelanotic lesion anywhere on the iris (**Fig. 8.36**).
- Anterior uveitis and pain are common.

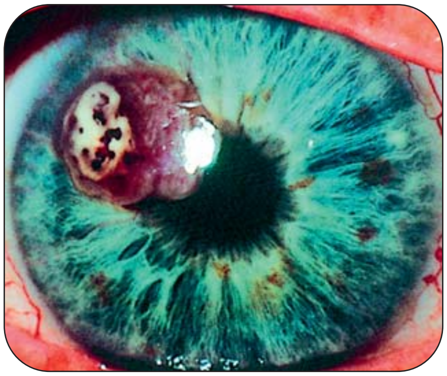

Fig. 8.36

Forward extension of ciliary body melanoma

Signs

- May mimic an iris melanoma (**Fig. 8.37**).
- Examination through a dilated pupil will reveal the ciliary body mass (**Fig. 8.38**).

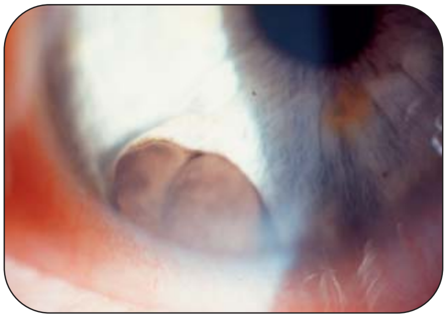

Fig. 8.37

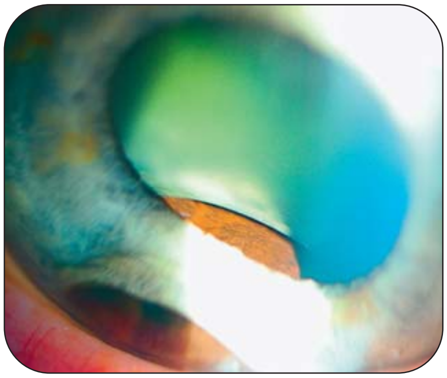

Fig. 8.38

Multiple

Freckles

Incidence

- Present in 50% of adults but rare before the age of 12 years.

Signs

- Bilateral spots ranging in colour from yellow-tan to deep chocolate-brown (**Fig. 8.39**).
- They are smaller than naevi and never distort the iris architecture.

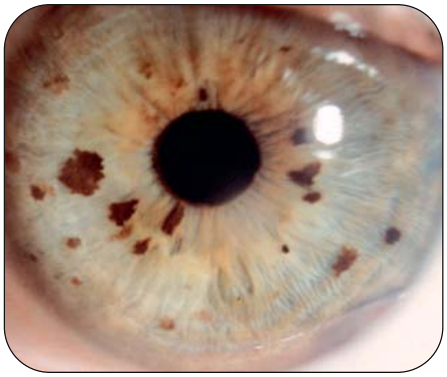

Fig. 8.39

Brushfield spots

Incidence

- Present in 85% of patients with Down syndrome and occasionally seen in normals.

Signs

- Bilateral white or yellowish spots arranged in a ring at the junction between the outer and middle third of the iris stroma (**Fig. 8.40**).

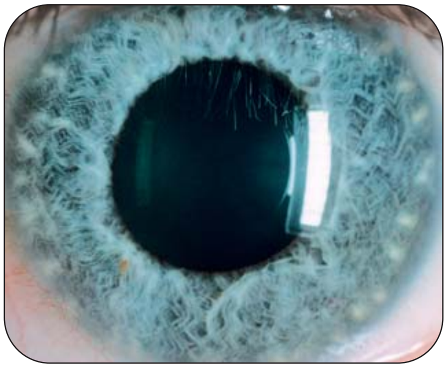

Fig. 8.40

Lisch nodules

Incidence

- Present in virtually all patients with NF1 after the age of 16 years.

Signs

- Bilateral yellow or brown dome-shaped nodules scattered throughout the stroma (**Fig. 8.41**).

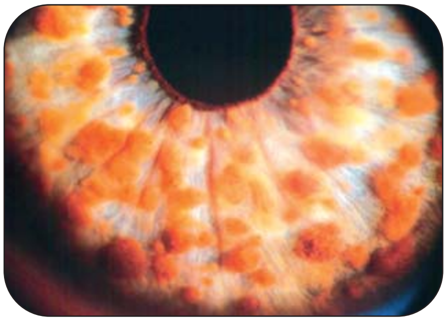

Fig. 8.41

Mammillations

Signs

- Multitude of bilateral regularly spaced villiform lesions (**Fig. 8.42**).

Look for

- Congenital ocular melanocytosis.
- NF1.
- Axenfeld–Rieger anomaly.
- Peters anomaly.

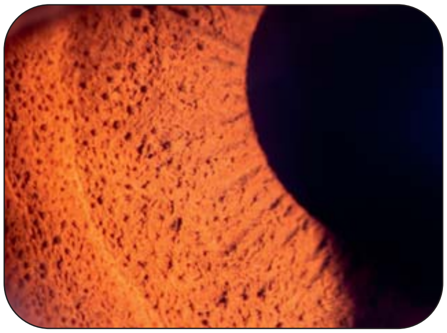

Fig. 8.42

Iris naevi in Cogan–Reese syndrome

Definition

- Cogan–Reese syndrome is part of the iridocorneal syndrome that typically affects middle-aged women.

Signs

- Unilateral, small, pedunculated nodules slightly larger than mammillations (**Fig. 8.43**).

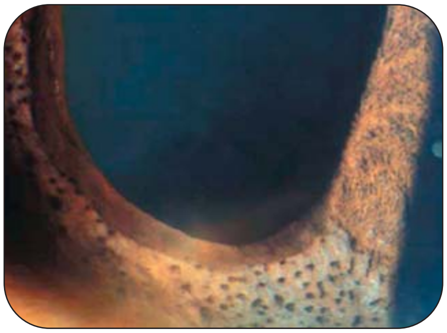

Fig. 8.43

Inflammatory nodules

Associations

- Nodules occur in GAU and Fuchs syndrome.

Signs

- Koeppe nodules are small and on the pupillary border (**Fig. 8.44**).
- Busacca nodules are larger and are located away from the pupil (**Fig. 8.45**).

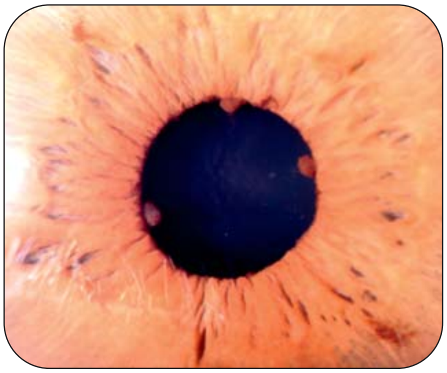

Fig. 8.44

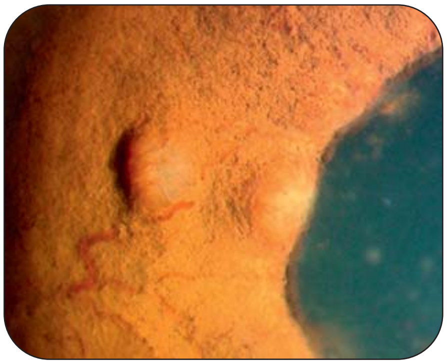

Fig. 8.45

Tapioca melanoma

Definition

- A very rare variant of iris melanoma.

Signs

- Lightly pigmented or non-pigmented nodules confined to one segment (**Fig. 8.46**).

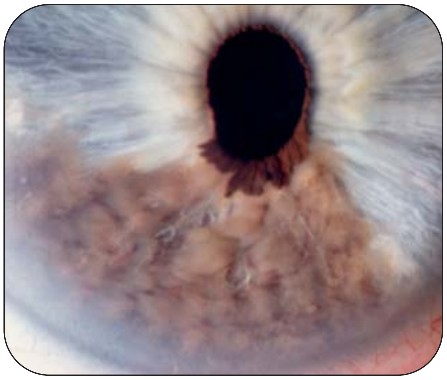

Fig. 8.46

Multiple iris metastases

Signs

- Pink or yellow fast-growing nodules (**Fig. 8.47**).
- Anterior uveitis and hyphaema may occur.

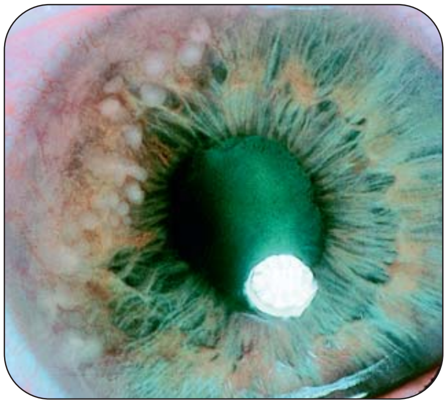

Fig. 8.47

Retinoblastoma invading the iris

Incidence

- Iris invasion is very rare and may give rise to diagnostic difficulties because it occurs in older children.

Signs

- Pale iris nodules (**Fig. 8.48**).
- Anterior uveitis and hyphaema may occur.

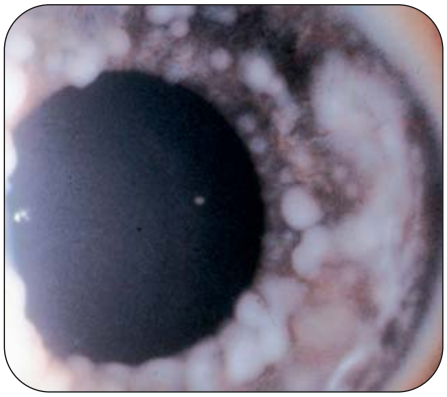

Fig. 8.48

Iris flocculi

Definition

- Uncommon congenital lesions.

Signs

- Pupillary tags or tiny solid nodules consisting of pigment epithelium (**Fig. 8.49**).

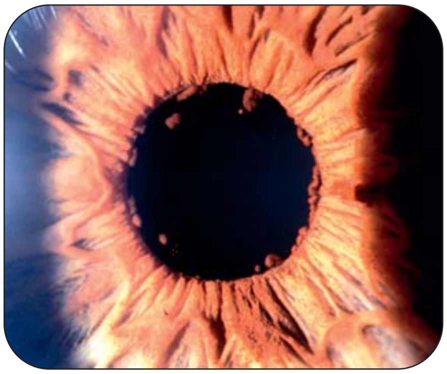

Fig. 8.49

CYSTIC LESIONS

Primary

Epithelial

Signs

- Unilateral or bilateral, and solitary or multiple.
- Dark-brown globular lesion that may be pupillary (**Fig. 8.50**), mid-zonal or peripheral (iridociliary – **Fig. 8.51**).

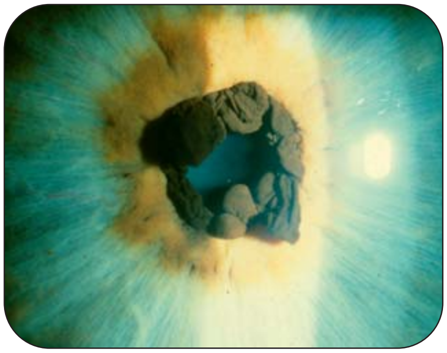

Fig. 8.50

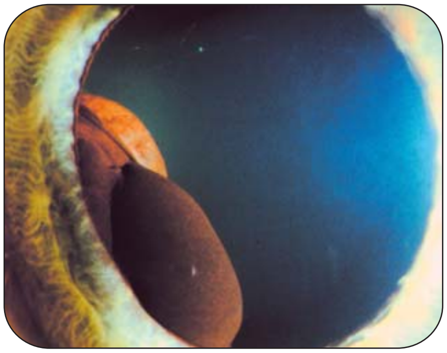

Fig. 8.51

Stromal

Signs

- Usually unilateral and solitary.
- The cyst contains fluid and has a clear anterior wall (**Fig. 8.52**).
- Sudden enlargement (**Fig. 8.53**) may cause glaucoma or endothelial damage.

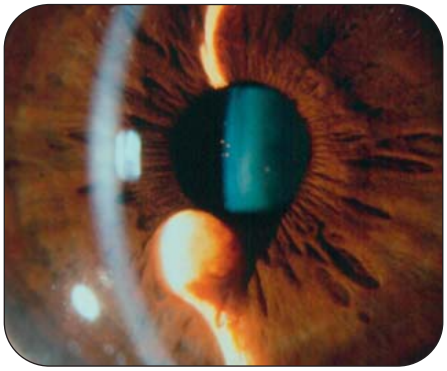

Fig. 8.52

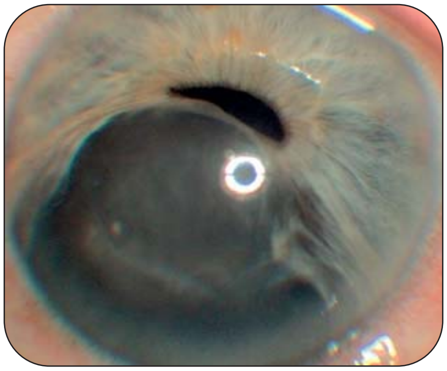

Fig. 8.53

Secondary

Implantation

Cause

- Epithelial down-growth following intraocular surgery or penetrating trauma.

Signs

- Similar to a primary stromal cyst but often connected in part to the wound (**Fig. 8.54**).

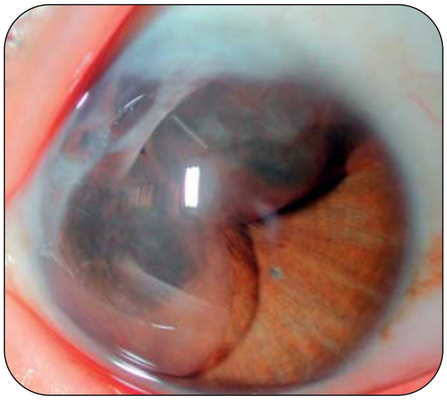

Fig. 8.54

Miotic

Cause

- Prolonged use of miotics, particularly ecothiopate iodide.

Signs

- Multiple, bilateral pupillary cysts (**Fig. 8.55**) similar to but smaller than primary pupillary epithelial cysts.

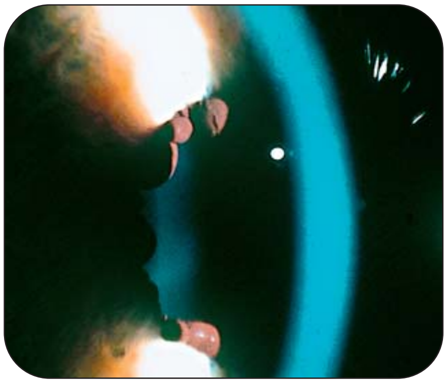

Fig. 8.55

Parasitic

Cause

- Parasitic cysts are extremely rare and are associated with encysted parasites such as in cysticercosis (**Fig. 8.56**).

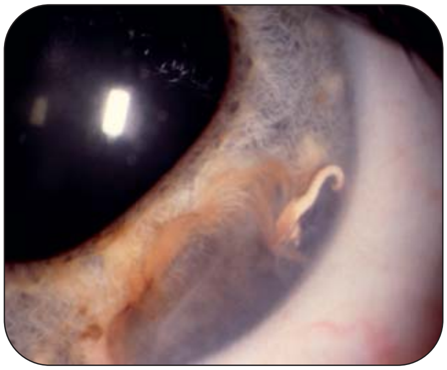

Fig. 8.56

ECTROPION UVEAE

Congenital

Signs

- Non-progressive condition with a round and reactive pupil (**Fig. 8.57**).
- May be associated with glaucoma due to an associated angle anomaly.

Associations

- NF1.
- Prader–Willi syndrome.
- Axenfeld–Rieger anomaly in which bilateral ectropion is associated with stromal hypoplasia (**Fig. 8.58**).

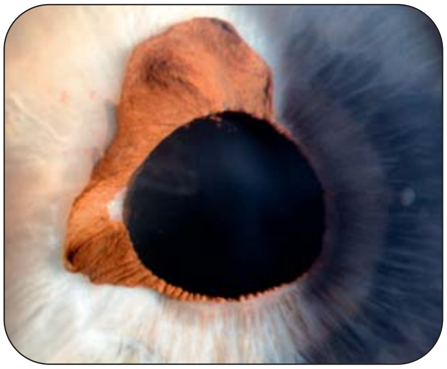

Fig. 8.57

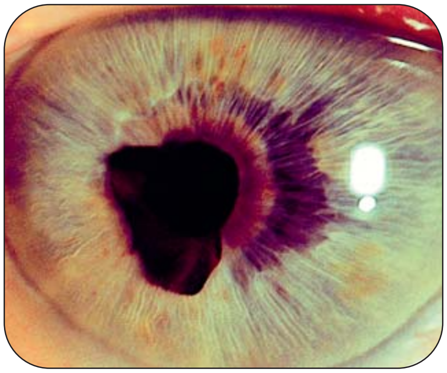

Fig. 8.58

Acquired

Associated with rubeosis iridis

Ectropion uveae occurs in end-stage disease and is usually mild (**Fig. 8.59**).

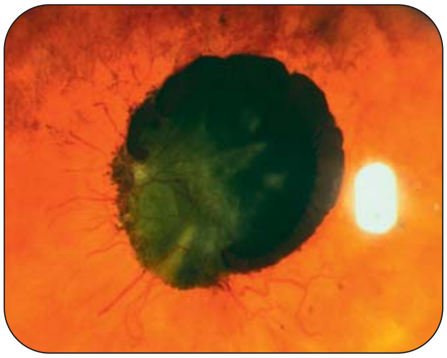

Fig. 8.59

Associated with iris tumours

Ectropion uveae is most frequently associated with melanoma (**Fig. 8.60**) and occasionally with large naevi.

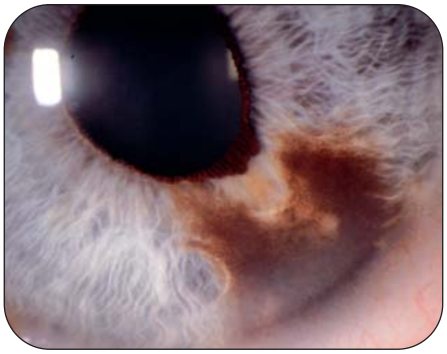

Fig. 8.60

RUBEOSIS IRIDIS

Signs

- Tiny dilated capillary tufts or red spots at the pupillary margins (**Fig. 8.61**).
- Radial surface iris neovascularisation (**Fig. 8.62**).

Causes

- Ischaemic central retinal vein occlusion.
- Diabetes mellitus.
- Chronic intraocular inflammation.
- Central retinal artery occlusion.
- Ocular ischaemic syndrome.
- Intraocular tumours.
- Long-standing retinal detachment.
- Retinopathy of prematurity.

Differential diagnosis

- Dilated iris capillaries (**Fig. 8.63**) associated with various types of anterior uveitis. When seen in patients with secondary syphilis they are referred to as roseolae.
- Iris haemangioma or vascular malformation (**Fig. 8.64**).
- Pupillary neovascular (Cobb) tufts are tiny lesions which may cause spontaneous hyphaema (**Fig. 8.65**).

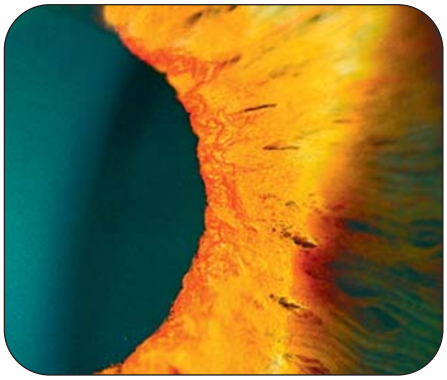

Fig. 8.61

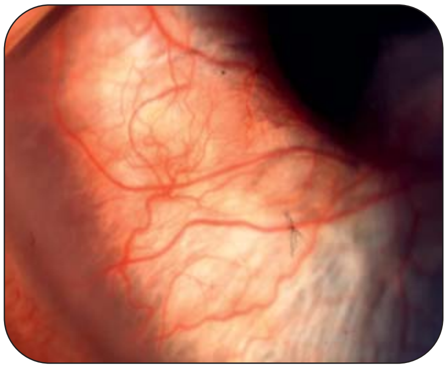

Fig. 8.62

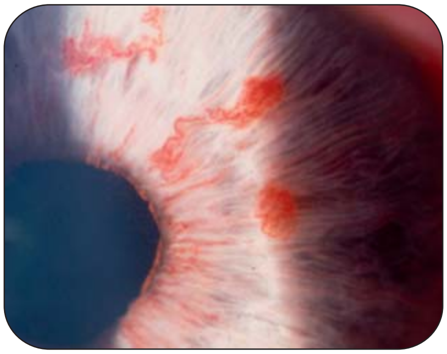

Fig. 8.63

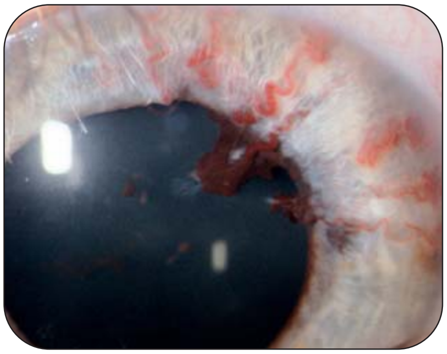

Fig. 8.64

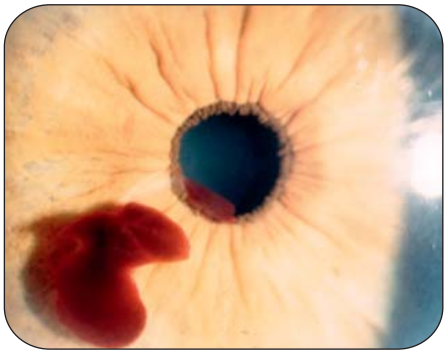

Fig. 8.65

IRIS ATROPHY AND TRANSLUCENCY

Fuchs syndrome

Signs

- Unilateral diffuse transillumination defect due to atrophy of the pigment epithelium (**Fig. 8.66**).

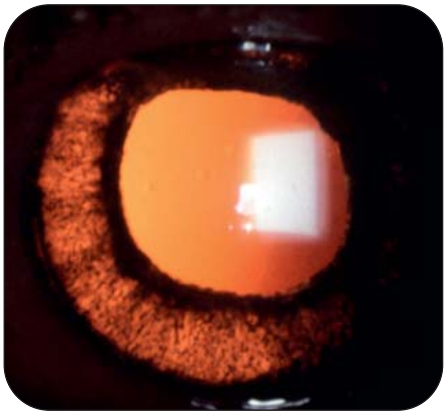

Fig. 8.66

Albinism

Classification

- Albinism may be ocular, in which only the eyes are involved, or oculocutaneous which involves the eyes, skin and hair (**Fig. 8.67**); the latter may be tyrosinase-negative or tyrosinase-positive.

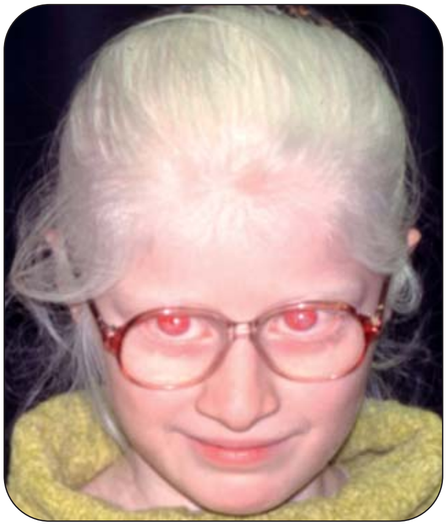

Fig. 8.67

Signs

a. *Tyrosinase-negative oculocutaneous albinism* manifests gross translucency of a blue iris with visibility of the edge of the lens and ciliary processes (**Fig. 8.68**).

b. *Tyrosinase-positive oculocutaneous albinism* shows milder translucency (**Fig. 8.69**).

c. *Ocular albinos* show variable translucency.

d. *Carriers* of X-linked ocular albinism may show mild translucency.

Look for

- Photophobia and nystagmus.
- Fundus hypopigmentation and foveal hypoplasia (**Fig. 8.70**).

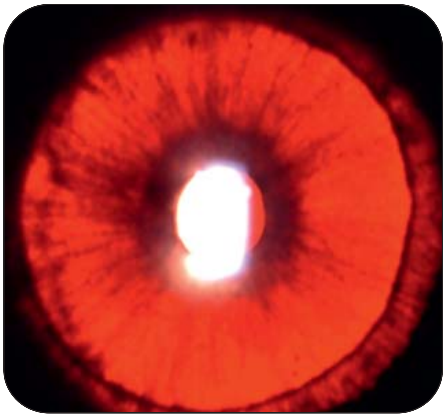

Fig. 8.68

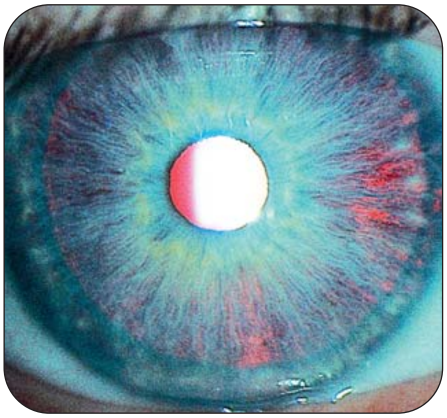

Fig. 8.69

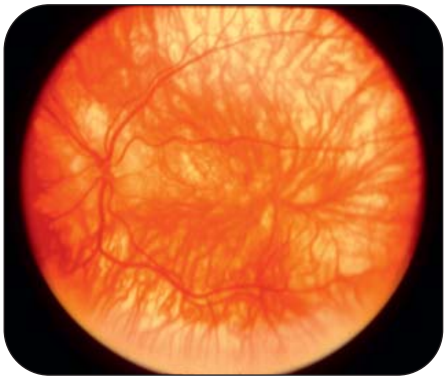

Fig. 8.70

- Chédiak–Higashi syndrome (recurrent infections).
- Hermansky–Pudlak syndrome (easy bruising).

Axenfeld–Rieger anomaly

Definition

- A rare, bilateral, non-progressive congenital condition.

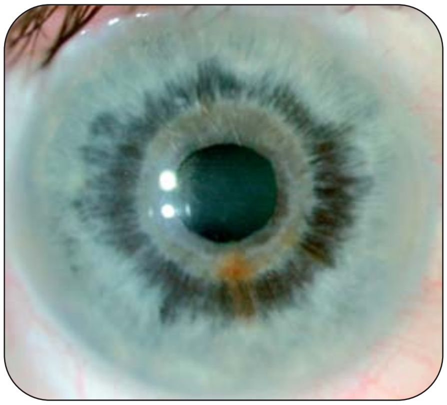

Fig. 8.71

Signs

- Diffuse hypoplasia of anterior iris stroma giving rise to prominence of the sphincter muscle (**Fig. 8.71**).

Look for ocular

- Posterior embryotoxon.
- Pseudopolycoria.
- Ectropion uveae (see Fig. 8.58).
- Corectopia.

Look for systemic (Rieger syndrome)

- Dental anomalies – microdontia and hypodontia (**Fig. 8.72**).
- Facial anomalies – maxillary hypoplasia (**Fig. 8.73**).
- Redundant paraumbilical skin (**Fig. 8.74**).
- Hypospadias.

Table 8.2 **Divisions of Axenfeld–Rieger anomaly and syndrome**

Condition	Angle anomalies	Iris anomalies	Systemic features	Glaucoma risk
Axenfeld anomaly	+	–	–	+
Rieger anomaly	++	+	–	+++
Rieger syndrome	++	+	+	+++

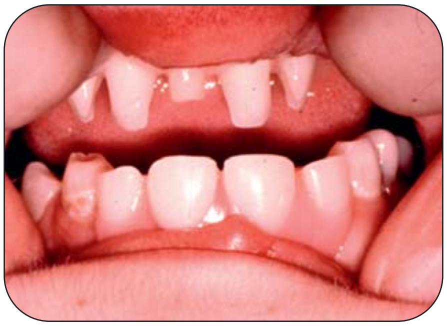

Fig. 8.72

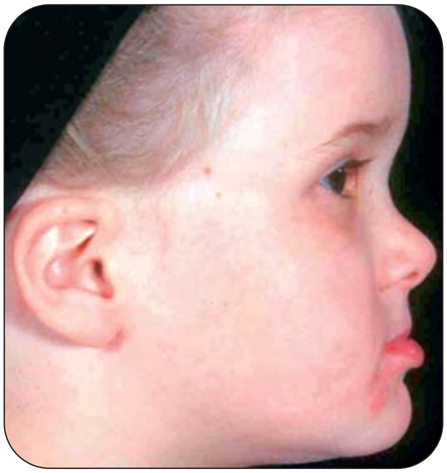

Fig. 8.73

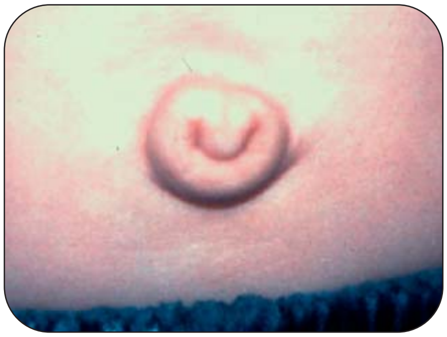

Fig. 8.74

Post acute angle-closure

Signs

- Stromal iris atrophy with a spiral-like configuration (**Fig. 8.75**).

Look for

- Fixed and semi-dilated pupil, shallow anterior chamber and narrow or closed angle.
- Intraocular pressure may be normal, subnormal or elevated.

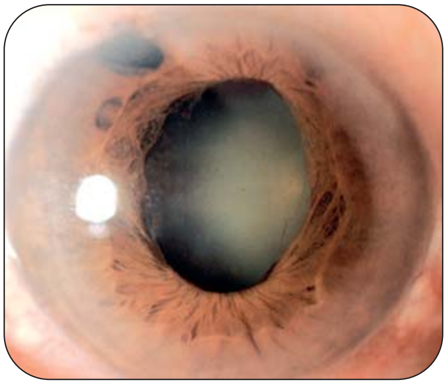
Fig. 8.75

Iridoschisis

Definition

- A rare, usually bilateral but asymmetric condition which typically affects elderly individuals.

Signs

- Intrastromal iris atrophy which is frequently inferior (**Fig. 8.76**).

Look for

- Shallow anterior chamber.
- Narrow angle and predisposition to angle closure; note peripheral iridotomy in Fig. 8.76.

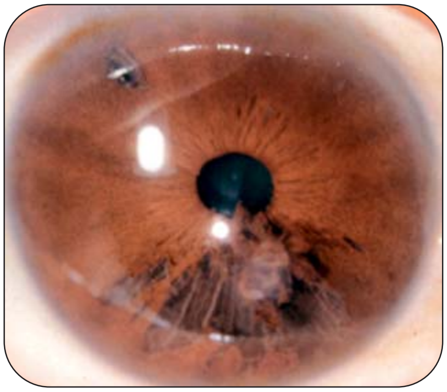
Fig. 8.76

Pigment dispersion syndrome

Definition

- An uncommon bilateral condition which typically affects young myopic men.

Signs

- Radial, mid-peripheral, slit-like transillumination defects (**Fig. 8.77**) are characteristic.

Look for

- Krukenberg spindle (see Fig. 7.4).
- Iris pigment granules.
- Deep anterior chamber.
- Trabecular hyperpigmentation (see Fig. 7.3).
- Glaucoma.

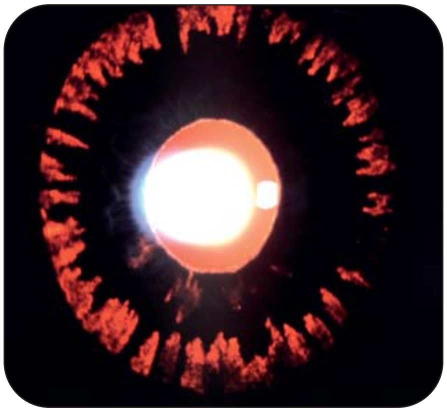
Fig. 8.77

Pseudophakic pigment dispersion

Signs

- Unilateral atrophic areas caused by rubbing of a posterior chamber intraocular lens against the back of the iris (**Fig. 8.78**).

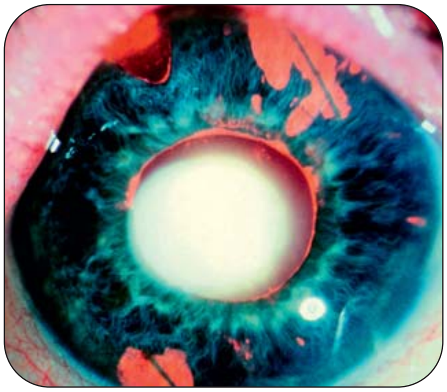

Fig. 8.78

Pseudoexfoliation syndrome

Definition

- An uncommon usually bilateral condition that affects the elderly.

Signs

- Subtle moth-eaten sphincter atrophy (**Fig. 8.79**).

Look for

- Pseudoexfoliation on the anterior lens surface (see Fig. 7.2).
- Trabecular hyperpigmentation (see Fig. 7.1).
- Increased risk of glaucoma.

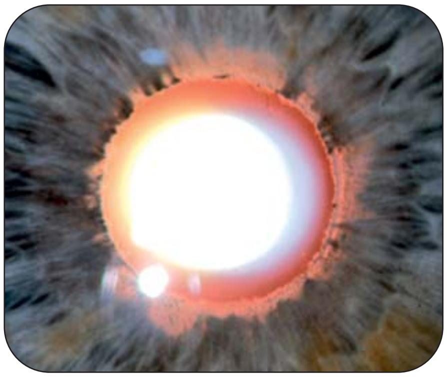

Fig. 8.79

Diabetic iridopathy

Signs

- Bilateral minute transillumination defects due to shedding of the pigment epithelium (**Fig. 8.80**).

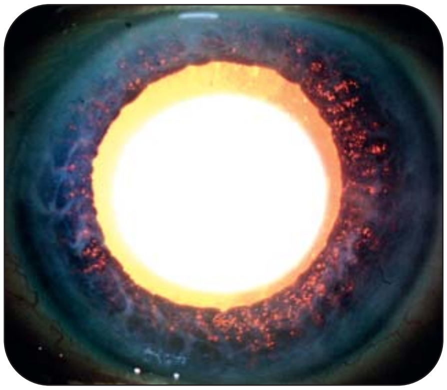

Fig. 8.80

Herpes zoster and simplex anterior uveitis

Signs

- Sectoral defects (**Fig. 8.81**).

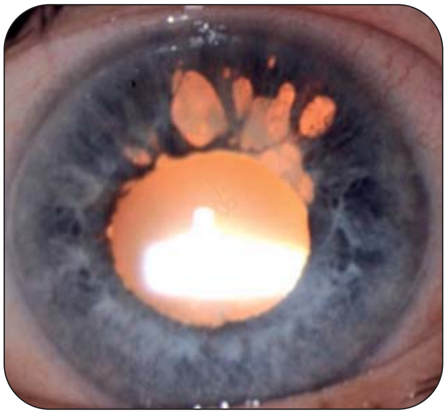

Fig. 8.81

Lepromatous iritis

Signs

- Severe miosis and slowly progressive iris atrophy that may result in full-thickness defects (**Fig. 8.82**).

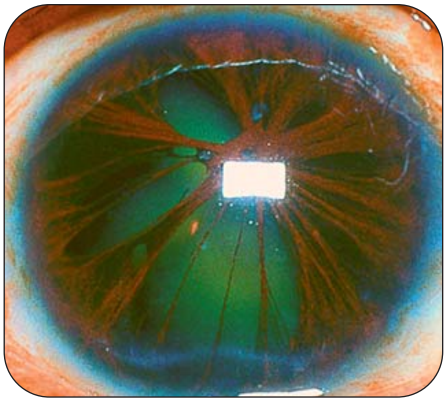

Fig. 8.82

Rubella iritis

Signs

- Severe diffuse iris atrophy (**Fig. 8.83**).

Look for

- Keratitis.
- Cataract.
- Microphthalmos.
- Retinopathy.

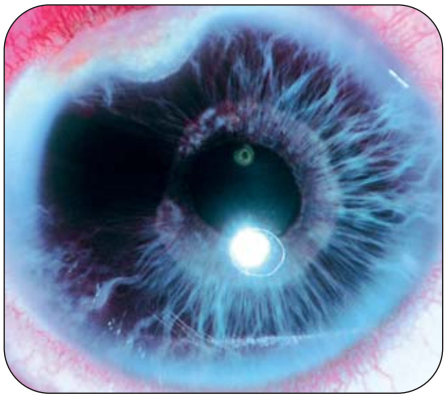

Fig. 8.83

FULL-THICKNESS DEFECT OR ABSENCE

Aniridia

Definition

- A rare bilateral congenital condition of which there are three types (Table 8.3).

Signs

- Absence of the iris may be partial (**Fig. 8.84**) or total (**Fig. 8.85**).

Look for

- Synechial angle-closure glaucoma as a result of pulling forward of rudimentary iris tissue (see Fig. 7.20).
- Associated systemic disorders.
- Associated ocular conditions (Table 8.4).

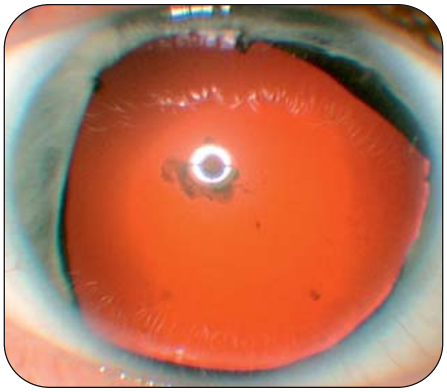

Fig. 8.84

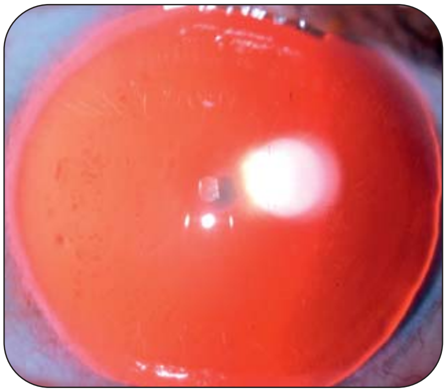

Fig. 8.85

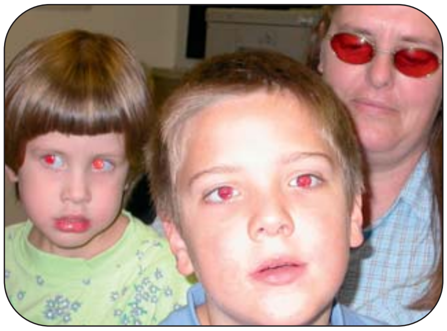

Fig. 8.86

Table 8.3 **Classification of aniridia**

1. **AN-1 (85%)**
   - Isolated (AD – **Fig. 8.86**)
2. **AN-2 – Miller syndrome (13%)**
   - Deletion of the short arm of chromosome 11
   - Wilms tumour
   - Genitourinary anomalies
   - Mental handicap
3. **AN-3 – Gillespie syndrome (2%)**
   - AR
   - Mental handicap
   - Cerebellar ataxia

Table 8.4 **Ocular associations of aniridia**

1. **Glaucoma**
2. **Cornea**
   - Tear film instability and epithelial defects
   - Limbal stem cell deficiency and pannus
   - Opacity
   - Dermoids
   - Microcornea
   - Sclerocornea
   - Keratolenticular adhesions
3. **Lens**
   - Absence
   - Anterior polar cataract
   - Subluxation (see Fig. 7.21)
   - Persistent pupillary membranes
4. **Fundus**
   - Foveal hypoplasia
   - Disc hypoplasia
   - Coloboma
5. **Nystagmus**

Iris coloboma

Pathogenesis

- An uncommon unilateral or bilateral congenital condition caused by defective closure of the embryonic fissure. Isolated iris colobomas are either sporadic or AD.

Signs

- Total coloboma involves the whole iris from pupil to root (**Fig. 8.87**).
- Partial coloboma does not involve the iris root (**Fig. 8.88**).

Look for

- Ocular and systemic associations (Table 8.5).

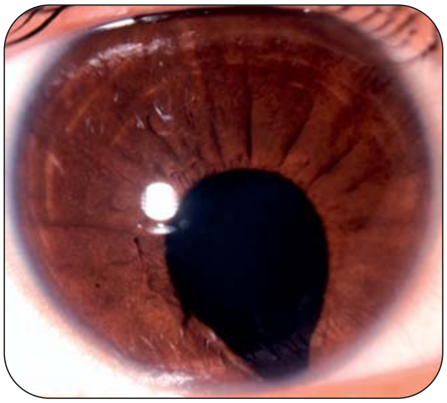

Fig. 8.87

Table 8.5 **Associations of iris coloboma**

1. **Ocular**
    - Microphthalmos (see Fig. 5.5)
    - Other colobomas
2. **Systemic**
    - Patau syndrome (trisomy 13)
    - Edward syndrome (trisomy 18)
    - Cat-eye syndrome (partial trisomy 22)
    - Pagon (CHARGE) syndrome
    - Meckel–Gruber syndrome
    - Lenz microphthalmos syndrome
    - Goltz focal dermal hypoplasia
    - Rubinstein–Taybi syndrome

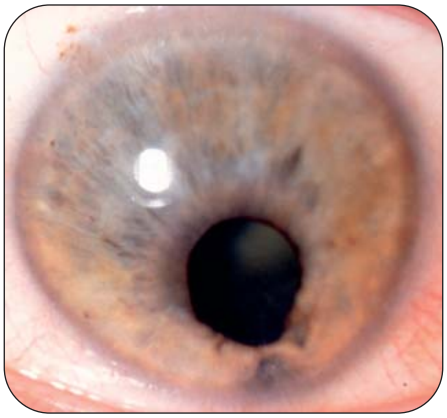

Fig. 8.88

Axenfeld–Rieger anomaly

Definition

- A rare bilateral, congenital condition.

Signs

- Non-progressive full-thickness iris defects (**Fig. 8.89**).

Look for

- Other anomalies (see above).

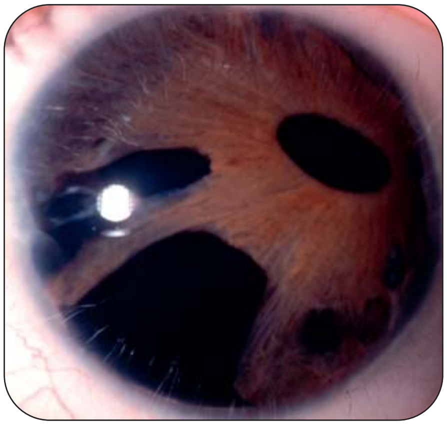

Fig. 8.89

Iridocorneal-endothelial syndrome

Definition

- A rare unilateral acquired condition that typically affects females.

Signs

- Full-thickness iris defects (pseudopolycoria – **Fig. 8.90**).

Look for

- Broad-based PAS.
- Glaucoma.
- Corectopia.

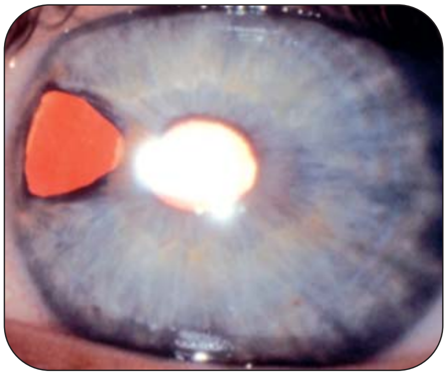

Fig. 8.90

Blunt trauma

Signs

- Iridodialysis (**Fig. 8.91**).

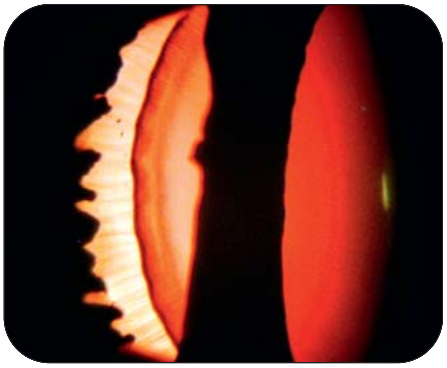

Fig. 8.91

Laser iridotomy

Signs

- Small superior defect (**Fig. 8.92**).

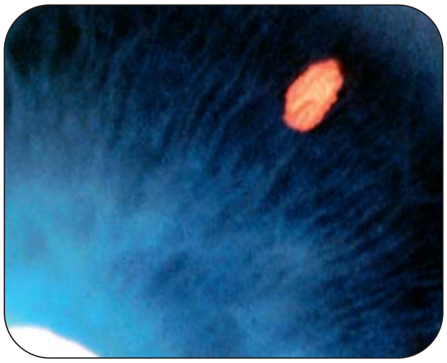

Fig. 8.92

HETEROCHROMIA IRIDIS

In heterochromia iridis the irides have different colours. The condition may be congenital or acquired and the affected iris may be hypochromic (lighter in colour) or hyperchromic (darker). Rarely the irides of both eyes have sectors with different colours.

Congenital

Isolated simple

Signs

- Hyperchromic heterochromia (**Fig. 8.93**).

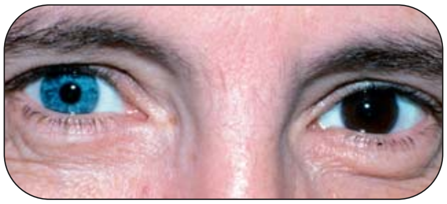

Fig. 8.93

Waardenburg syndrome

Definition

- A very rare AD condition.

Signs

- Hyperchromic heterochromia (**Fig. 8.94**).

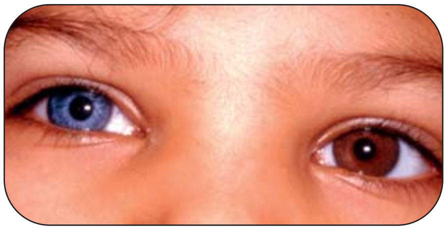

Fig. 8.94

Look for

- Partial albinism.
- Telecanthus.
- Poliosis and white hair forelock.
- Synophrys or unusual facial hair distribution.

Parry–Romberg syndrome

Definition

- A very rare idiopathic condition characterised by progressive hemifacial atrophy (**Fig. 8.95**).

Signs

- Hypochromic heterochromia is uncommon.

Look for

- Poliosis.
- Enophthalmos.
- Restrictive strabismus.
- Horner syndrome.

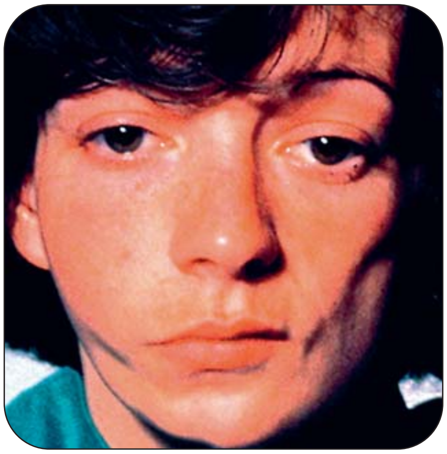

Fig. 8.95

Horner syndrome

Signs

- Ipsilateral hypochromic heterochromia may be seen in congenital lesions (**Fig. 8.96**) and occasionally if the lesion is acquired and long-standing.

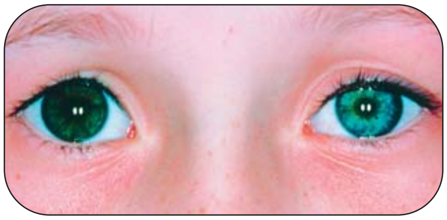

Fig. 8.96

Look for

- Miosis.
- Mild ptosis and elevation of the lower eyelid.
- Anhidrosis if the lesion is below the superior cervical ganglion.

Oculodermal melanocytosis (naevus of Ota)

Signs

- Hyperchromic heterochromia (**Fig. 8.97**) ipsilateral to cutaneous hyperpigmentation is common.

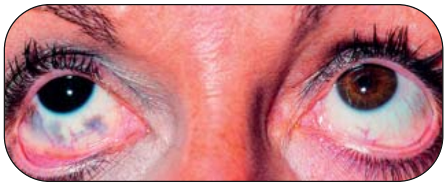

Fig. 8.97

Look for

- Episcleral hyperpigmentation.
- Trabecular and fundus hyperpigmentation.
- Uveal melanoma.
- Glaucoma.

Sturge–Weber syndrome

Signs

- Hyperchromic heterochromia ipsilateral to the naevus flammeus is uncommon.

Look for

- Episcleral haemangioma.
- Diffuse choroidal haemangioma.
- Glaucoma.
- Leptomeningeal haemangioma.

Hirschsprung disease

Definition

- A rare disease characterised by congenital absence of intestinal ganglion cells resulting in constipation and abdominal distension (congenital megacolon).

Signs

- Bilateral sectoral hypochromia (**Fig. 8.98**).

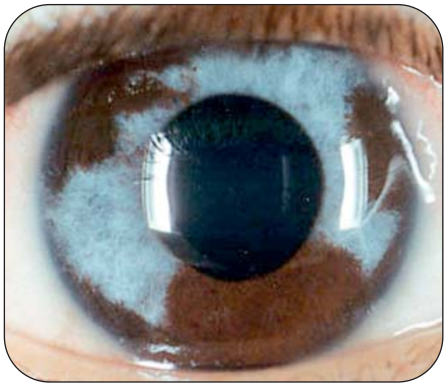

Fig. 8.98

Acquired

Fuchs syndrome

Signs

- Usually hypochromic heterochromia (**Fig. 8.99**) but in 10% of cases it may be hyperchromic (paradoxical).

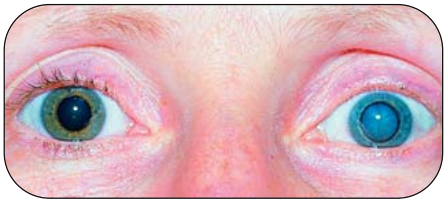

Fig. 8.99

Unilateral use of a prostaglandin analogue

Signs

- Hyperchromic heterochromia (**Fig. 8.100**), particularly in patients with green-brown or blue-brown irides.

Look for

- Lash lengthening and hyperpigmentation (see Fig. 1.17).
- Hyperpigmentation of periorbital skin.

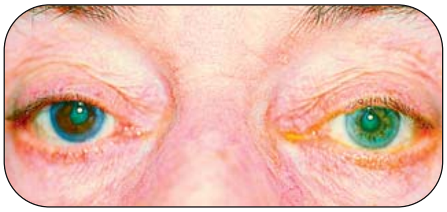

Fig. 8.100

Ocular siderosis

Signs

- Hyperchromic heterochromia (**Fig. 8.101**) due to reddish-brown iron deposition within the dilator muscle.

Look for

- Cataract.
- Glaucoma.
- Retinal degeneration (see Fig. 13.281).

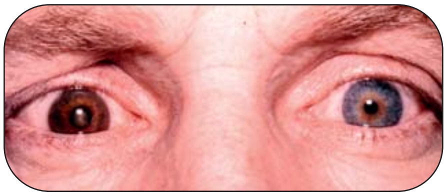

Fig. 8.101

Diffuse pigmented iris tumours

Signs

- Large naevus (**Fig. 8.102**) or melanoma.

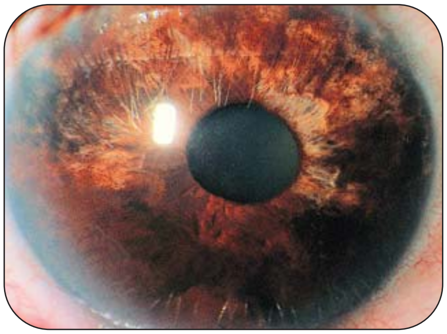

Fig. 8.102

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Pupils 9

ABNORMAL PUPILLARY REACTIONS **195**
- *Right physiological anisocoria* 195
- *Right pharmacological mydriasis* 196
- *Right episodic mydriasis* 197
- *Right tonic (Adie) pupil* 198
- *Right Horner syndrome* 199
- *Right third nerve palsy* 200
- *Argyll Robertson pupils* 201
- *Tectal (dorsal midbrain) pupils* 202
- *Right traumatic iridoplegia* 203

ABNORMAL PUPIL SIZE **204**

Small **204**
- *Anterior uveitis* 204
- *Spasm of the near reflex* 204
- *Argyll Robertson pupils* 204
- *Lepromatous miosis* 204
- *Congenital microcoria* 205
- *Ectopia lentis et pupillae* 205
- *Other causes* 205

Large **205**

WHITE PUPIL IN CHILDHOOD (LEUKOCORIA) **206**

Congenital **206**
- *Cataract* 206
- *Persistent anterior fetal vasculature* 206
- *Norrie disease* 206
- *Incontinentia pigmenti (Bloch–Sulzberger syndrome)* 207
- *Extensive white fundus lesions* 207

Acquired **208**
- *Retinoblastoma* 208
- *Cicatricial retinopathy of prematurity* 208
- *Chronic toxocaral endophthalmitis* 209
- *Coats disease* 209

ABNORMAL PUPILLARY REACTIONS

Right physiological anisocoria

Signs (Fig. 9.1)

a. In dim light the right pupil is larger than the left.

b. In bright light both pupils constrict normally.

c. After instillation of cocaine 10% to both eyes, both pupils dilate.

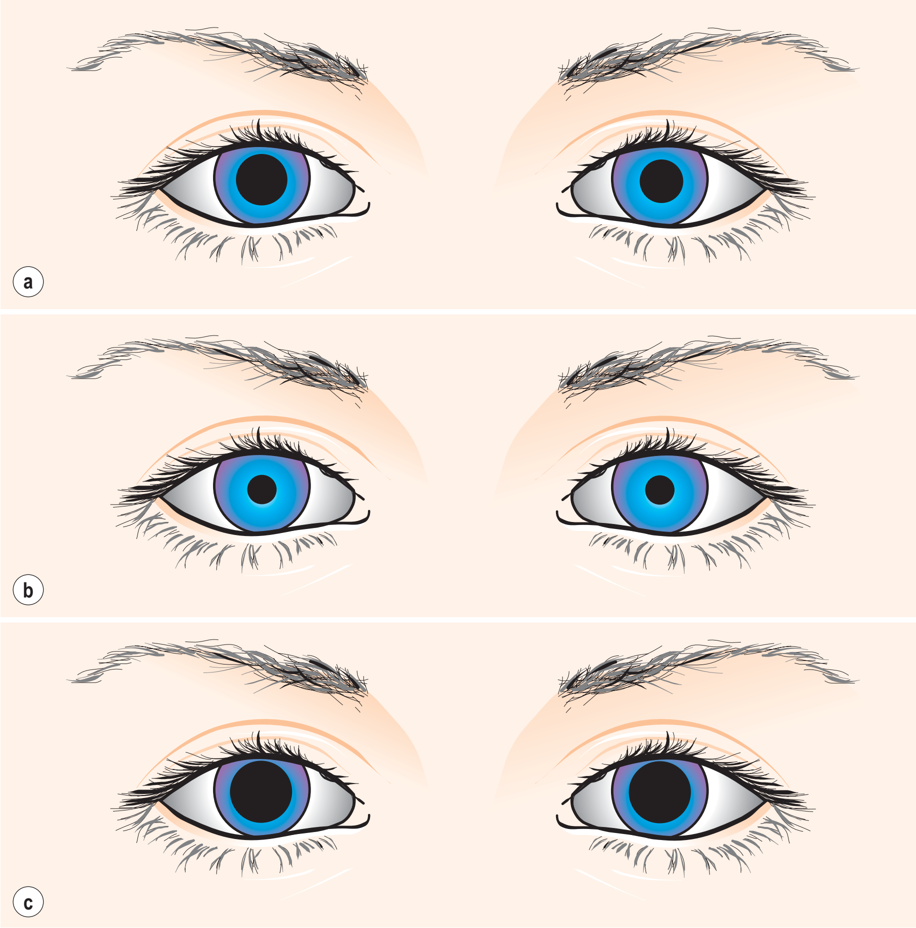

Fig. 9.1

Right pharmacological mydriasis

Signs (Fig. 9.2)

a. In dim light there is right mydriasis.

b. In bright light the right pupil does not constrict.

c. On accommodation the right pupil does not constrict.

d. After instillation of pilocarpine 1/10% to both eyes neither pupil constricts.

e. After instillation of pilocarpine 1% to both eyes, the right pupil does not constrict but the left does.

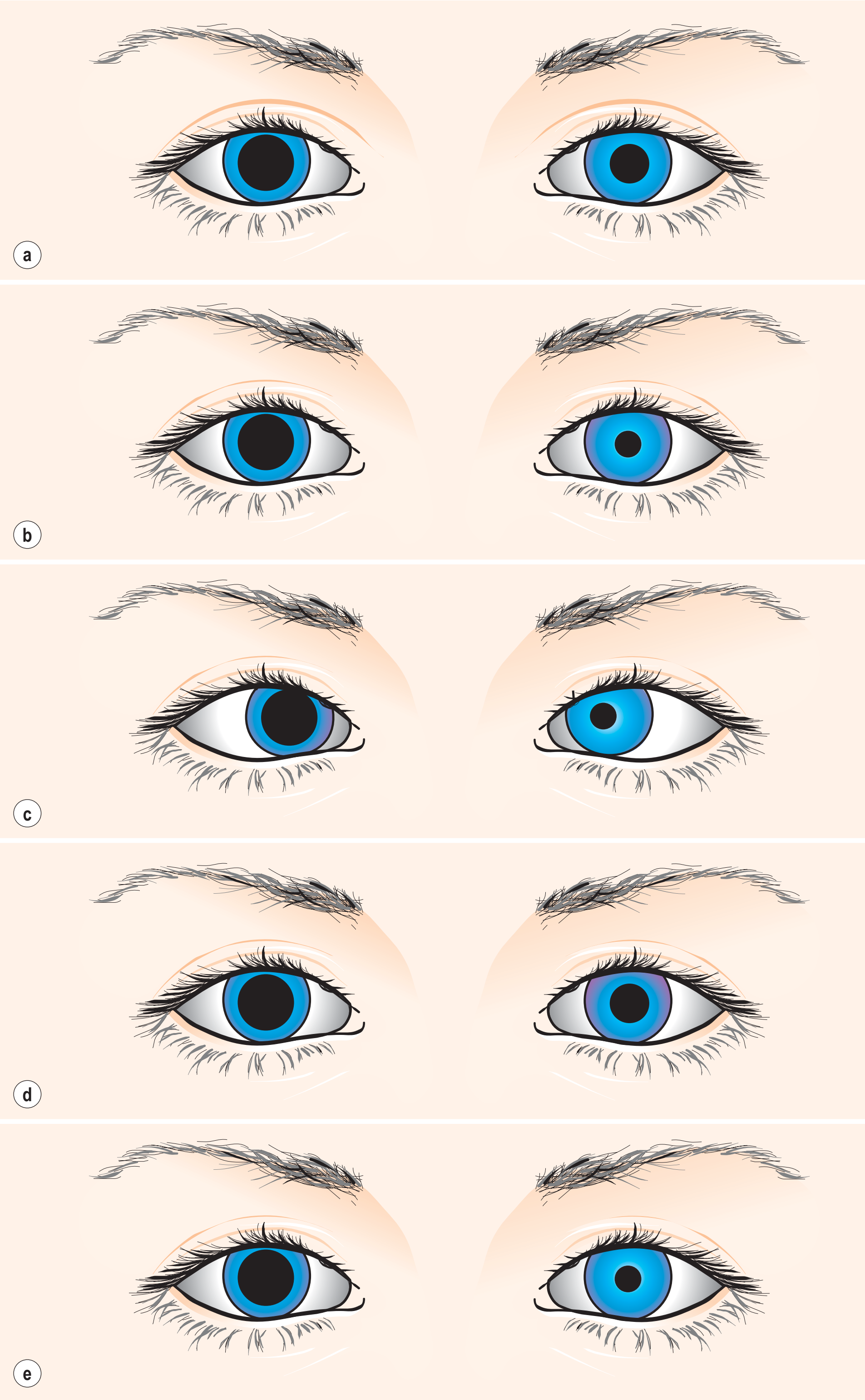

Fig. 9.2

Right episodic mydriasis

Signs (Fig. 9.3)

a. In dim light the right pupil is larger than the left.

b. In bright light the right pupil does not constrict.

c. On accommodation the right pupil does not constrict.

d. After instillation of pilocarpine 1/10% to both eyes, neither pupil constricts.

e. After instillation of pilocarpine 1% to both eyes, both pupils constrict.

f. After 24 hours both pupils are of equal size.

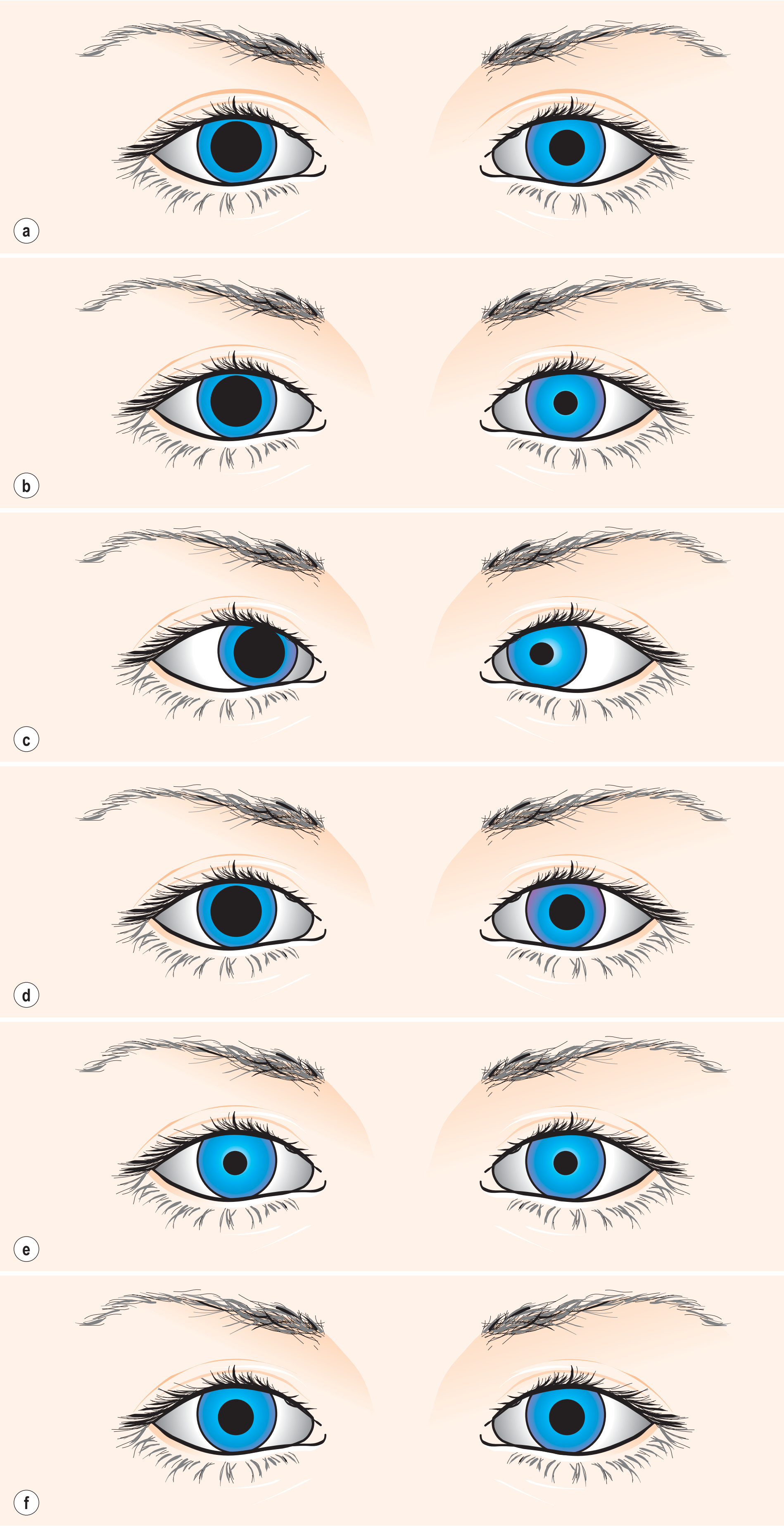

Fig. 9.3

Right tonic (Adie) pupil

Signs (Fig. 9.4)

a. In dim light there is right mydriasis.

b. In bright light there is right mydriasis.

c. On accommodation both pupils constrict although the right constricts very slowly and segmentally.

d. After instillation of pilocarpine 1/10% to both eyes, the right pupil constricts but not the left.

Look for

- Diminished or absent deep tendon reflexes (Holmes–Adie syndrome).

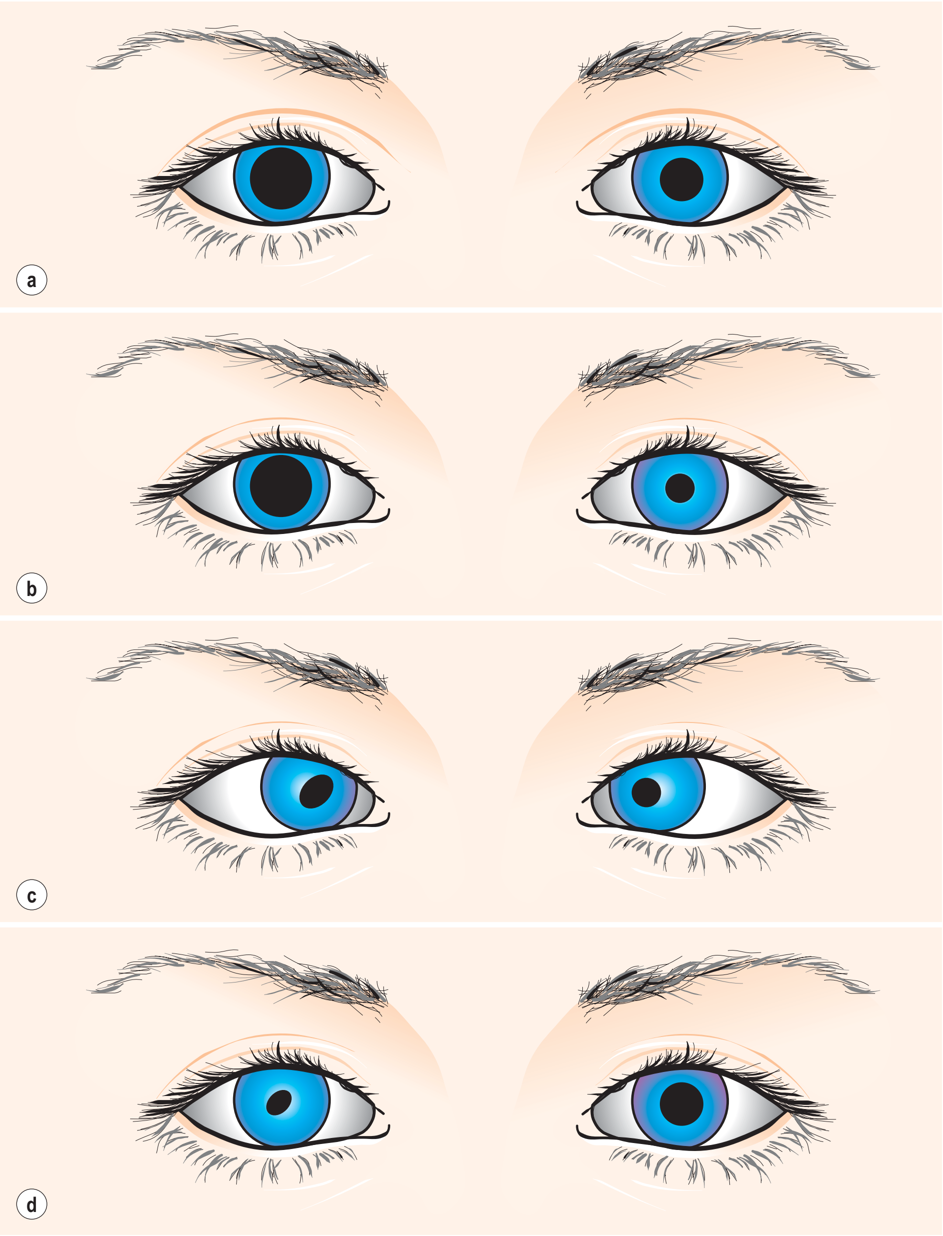

Fig. 9.4

Right Horner syndrome

Signs (Fig. 9.5)

a. In dim light there is right miosis and mild ptosis.

b. In bright light both pupils constrict normally.

c. After instillation of cocaine 10% to both eyes, the right pupil dilates less than the left.

Look for

- Elevation of lower lid.
- Heterochromia iridis (if congenital or long-standing – see Fig. 8.96).
- Anhidrosis if the lesion is below the superior cervical ganglion.

Causes

- See Table 1.7.

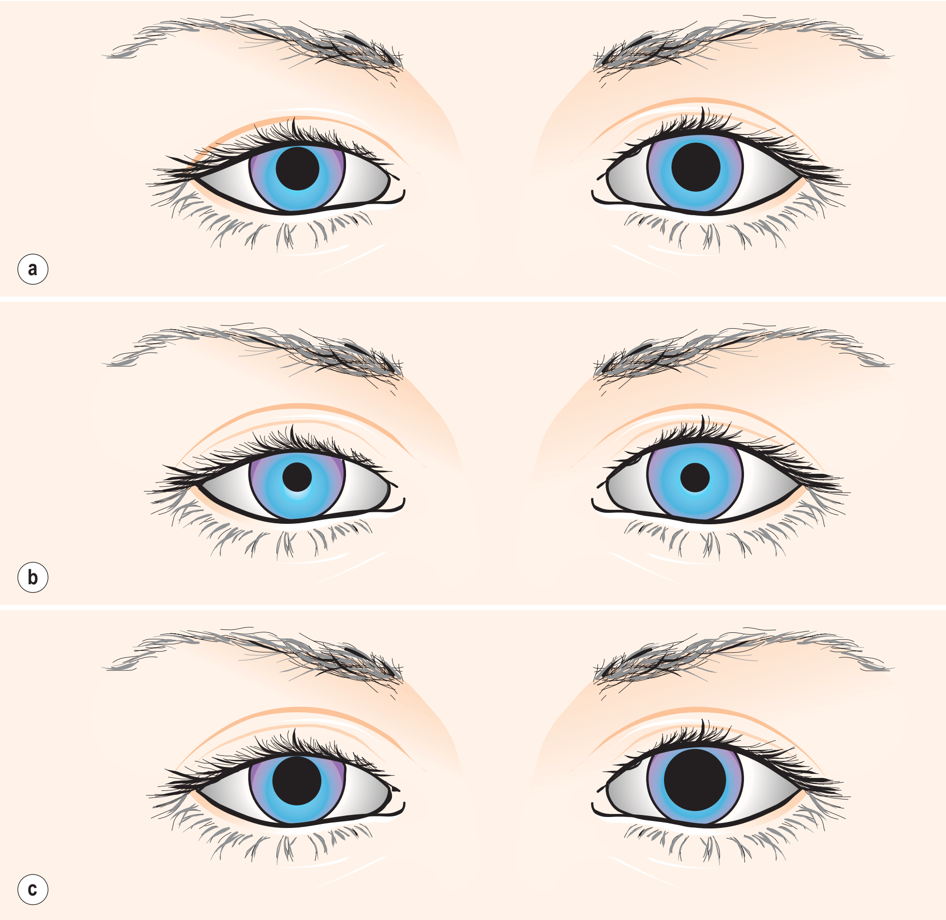

Fig. 9.5

Right third nerve palsy

Signs (Fig. 9.6)

a. In dim light there is right mydriasis associated with ptosis and ophthalmoplegia.

b. In bright light the right pupil does not constrict.

c. On accommodation the right pupil does not constrict.

d. After instillation of pilocarpine 1/10% into both eyes, neither pupil constricts.

e. After instillation of pilocarpine 1% into both eyes, both pupils constrict.

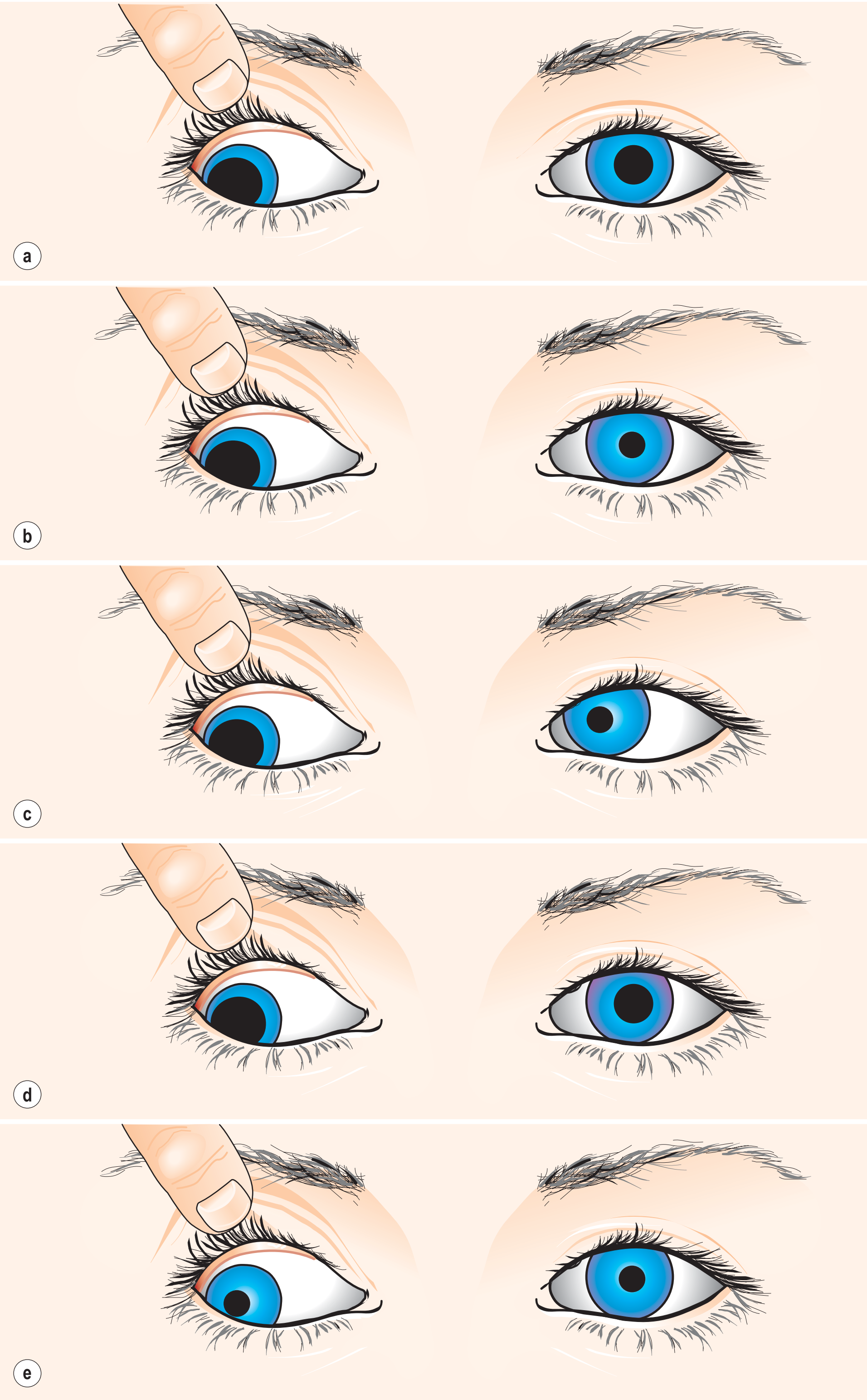

Fig. 9.6

Argyll Robertson pupils

Signs (Fig. 9.7)

a. In dim light both pupils are small.

b. In bright light neither pupil constricts.

c. On accommodation both pupils constrict (light-near dissociation – see Table 9.1).

d. After instillation of pilocarpine 1/10% to both eyes, neither pupil constricts.

Table 9.1 **Causes of light-near dissociation**

1. **Unilateral**
   - Afferent conduction defect
   - Herpes zoster ophthalmicus
   - Aberrant 3rd nerve regeneration
2. **Bilateral**
   - Juvenile-onset diabetes
   - Myotonic dystrophy
   - Parinaud dorsal midbrain syndrome (see Table 1.6)
   - Argyll Robertson pupils
   - Pituitary tumour
   - Familial amyloidosis
   - Encephalitis
   - Chronic alcoholism

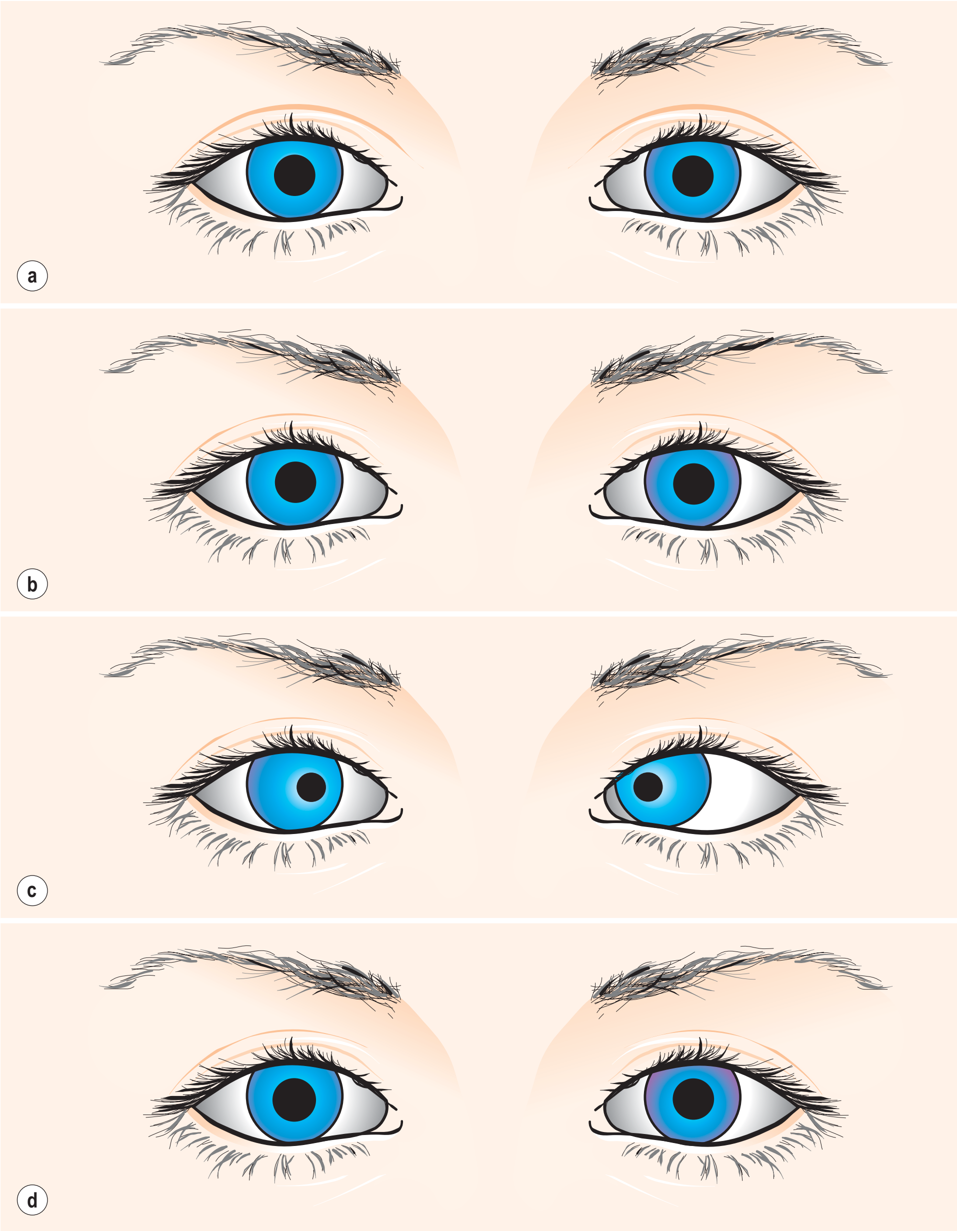

Fig. 9.7

Tectal (dorsal midbrain) pupils

Signs (Fig. 9.8)

a. In dim light there is bilateral asymmetric mydriasis.

b. In bright light neither pupil constricts.

c. On accommodation both pupils constrict normally.

d. After instillation of pilocarpine 1/10% to both eyes, neither pupil constricts.

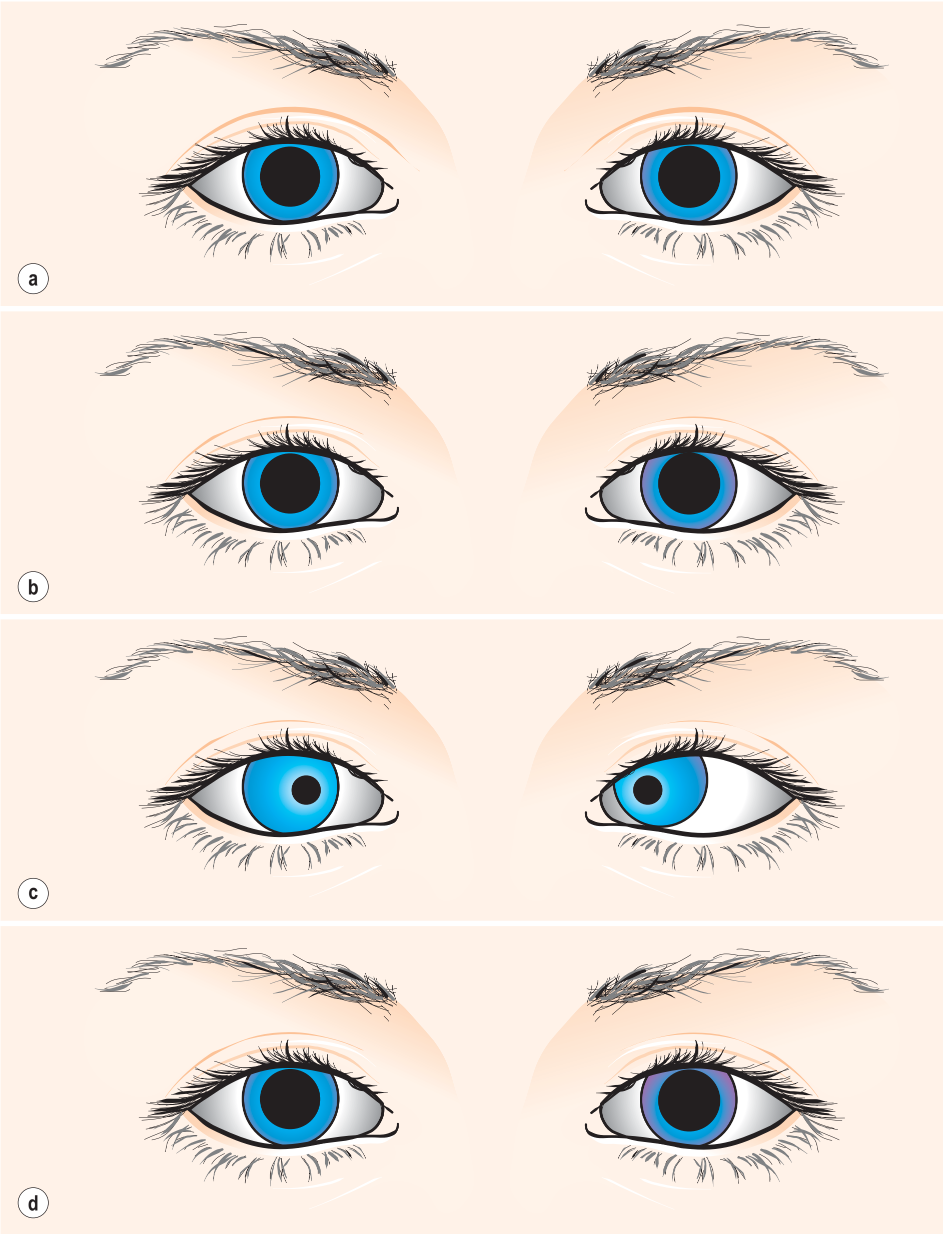

Fig. 9.8

Right traumatic iridoplegia

Signs **(Fig. 9.9)**

a. In dim light there is right mydriasis and the pupil is irregular in shape due to segmental damage.

b. In bright light the right pupil does not constrict.

c. On accommodation the right pupil does not constrict.

d. After instillation of pilocarpine 1/10% to both eyes, neither pupil constricts.

e. After instillation of pilocarpine 1% to both eyes, the right pupil constricts partially and segmentally, and the left normally.

Look for

- Sphincter tears and iridodialysis (see Fig. 8.91).

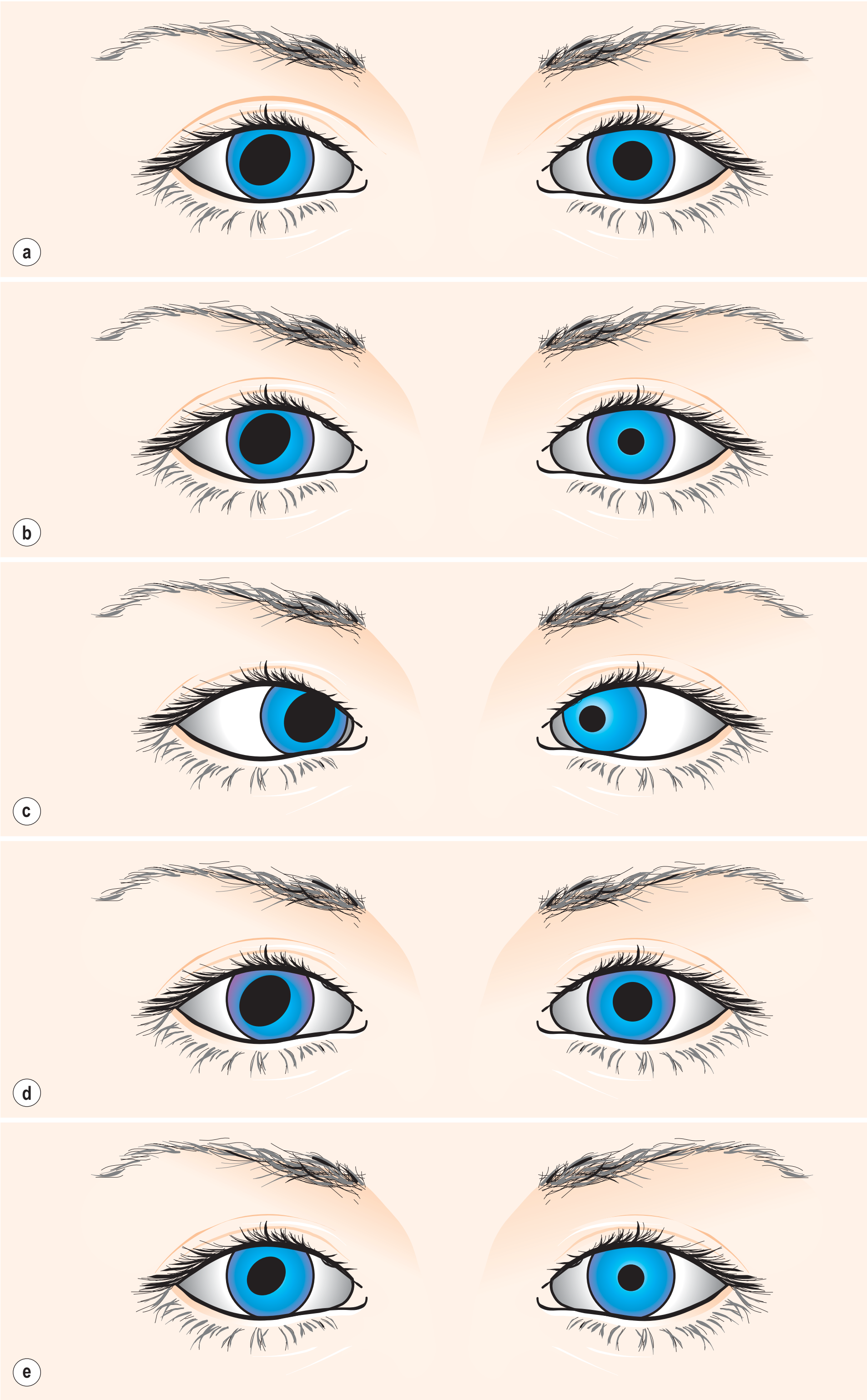

Fig. 9.9

ABNORMAL PUPIL SIZE

Small

Anterior uveitis

Signs

- In acute iritis miosis is caused by pupillary spasm (**Fig. 9.10**).
- In chronic iritis miosis may be caused by posterior synechiae (**Fig. 9.11**).

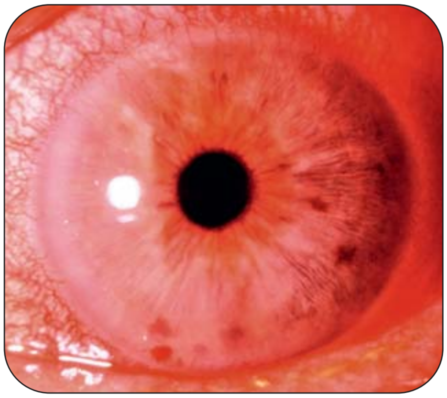

Fig. 9.10

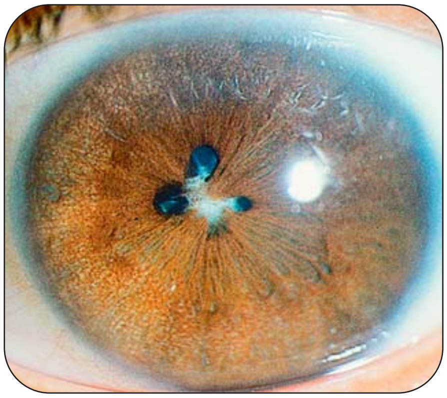

Fig. 9.11

Spasm of the near reflex

Signs

- Transient bilateral miosis, esotropia and myopia (**Fig. 9.12**).

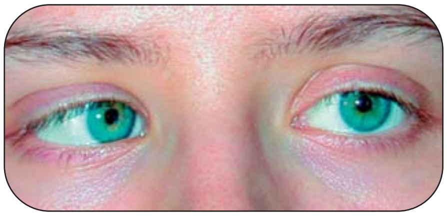

Fig. 9.12

Argyll Robertson pupils

Signs

- Bilateral miosis and frequently irregular-shaped pupils (**Fig. 9.13**)
- Light-near dissociation (see Fig. 9.7).

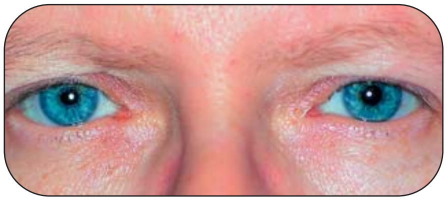

Fig. 9.13

Lepromatous miosis

Signs

- Bilateral miosis may occur due to damage to the sympathetic innervation to the sphincter pupillae in the absence of posterior synechiae (**Fig. 9.14**).

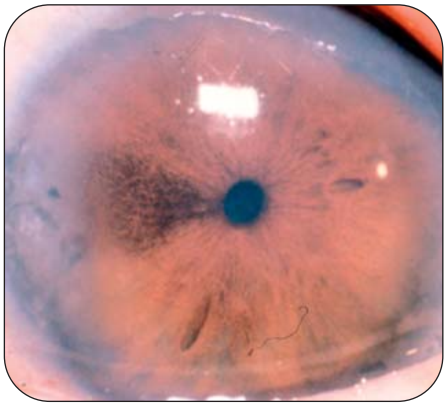

Fig. 9.14

Congenital microcoria

Signs

- Bilateral miosis and corectopia (**Fig. 9.15**).

Look for

- Shallow anterior chamber.
- Myopia.
- Glaucoma.

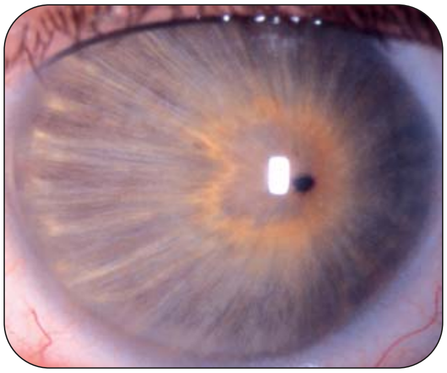

Fig. 9.15

Ectopia lentis et pupillae

Signs

- Bilateral miosis and corectopia with subluxation of the lens in the opposite direction (**Fig. 9.16**).

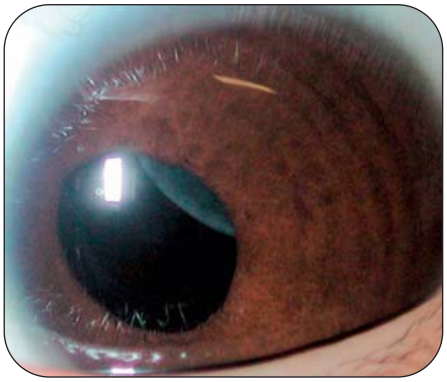

Fig. 9.16

Other causes

a. *Horner syndrome* (see above).

b. *Long-standing Adie pupil* ('little old Adie').

c. *Morphine.*

Large

Table 9.2 **Causes of large pupil**

- Adie pupil (see above)
- Third nerve palsy (see above)
- Uncal herniation
- Traumatic sphincter paralysis (see above)
- Iris ischaemia usually associated with iris atrophy (see Table 9.3)
- Postsurgical (Urrets-Zavalia syndrome) following penetrating keratoplasty for keratoconus
- Ocular siderosis
- Parinaud dorsal midbrain syndrome (see Table 1.6)
- Episodic mydriasis (see above)
- Brainstem death

Table 9.3 **Causes of iris ischaemia**

1. **Acute angle closure**
2. **Anterior segment ischaemia**
   - Squint surgery involving several muscles
   - Retinal detachment surgery with encirclement
   - Carotid artery disease

WHITE PUPIL IN CHILDHOOD (LEUKOCORIA)

Congenital

Cataract

Signs

- Dense unilateral or bilateral opacity (**Fig. 9.17**).

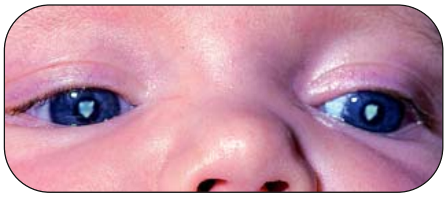

Fig. 9.17

Persistent anterior fetal vasculature

Definition

- A rare unilateral condition associated with mild microphthalmos (**Fig. 9.18**).

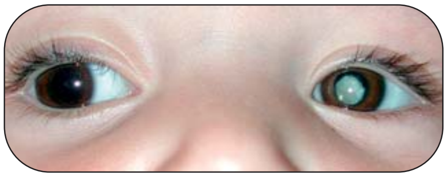

Fig. 9.18

Signs

- Elongated ciliary processes inserted into a retrolental mass (**Fig. 9.19**).

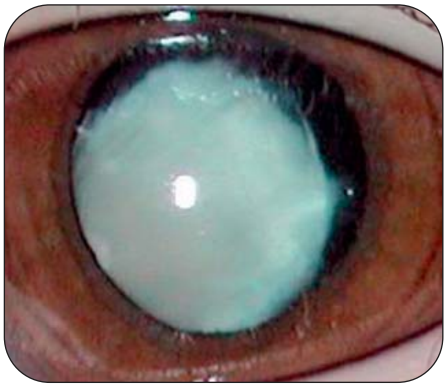

Fig. 9.19

Norrie disease

Definition

- An extremely rare X-LR condition characterised by developmental delay, deafness and perinatal blindness due to bilateral vitreoretinal dysplasia.

Signs

- Retinal folds and detachment.
- White retrolental masses (**Fig. 9.20**).
- Microphthalmos.
- Posterior synechiae.
- Shallow anterior chamber.

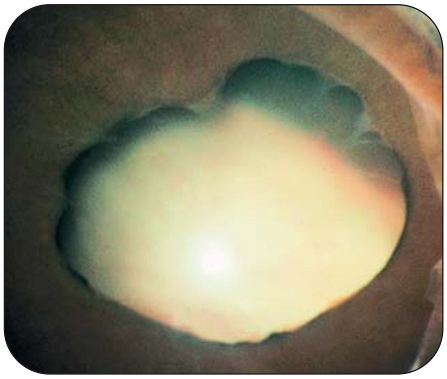

Fig. 9.20

Incontinentia pigmenti (Bloch–Sulzberger syndrome)

Definition

- An extremely rare X-LD disorder that manifests skin, dental and ocular abnormalities.
- Cutaneous bullae appear soon after birth (**Fig. 9.21**) and then slowly resolve to leave a linear pattern of hyperpigmentation (**Fig. 9.22**).

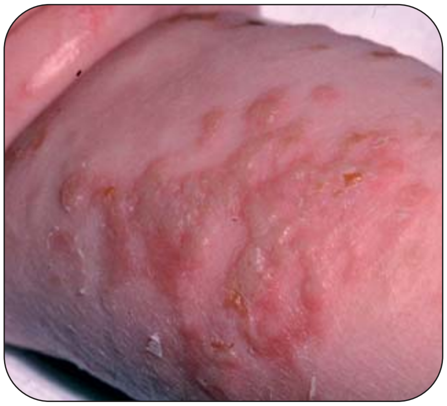

Fig. 9.21

Signs

- Leukocoria may be unilateral (**Fig. 9.23**) or bilateral.
- Retinal vascular abnormalities most marked in the temporal periphery.
- Total tractional retinal detachment in a minority of cases (**Fig. 9.24**).

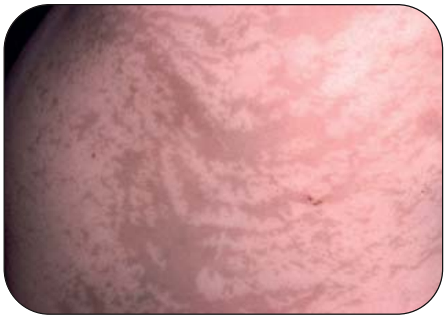

Fig. 9.22

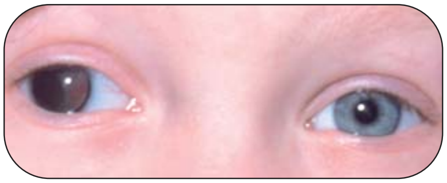

Fig. 9.23

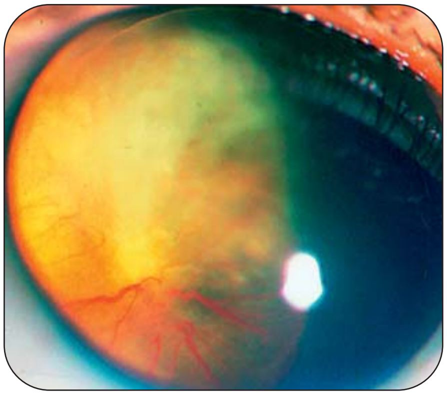

Fig. 9.24

Extensive white fundus lesions (Fig. 9.25)

a. *Myelination of nerve fibres* (**Fig. 9.26**).

b. *Chorioretinal colobomas* (**Fig. 9.27**).

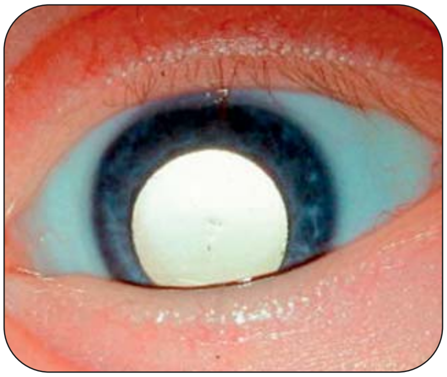

Fig. 9.25

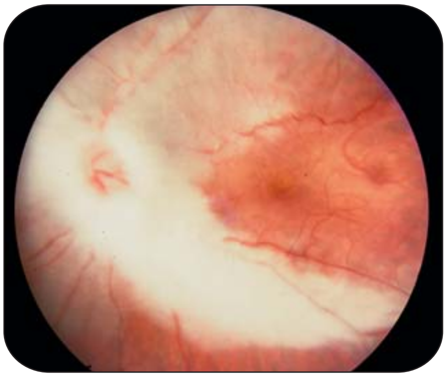

Fig. 9.26

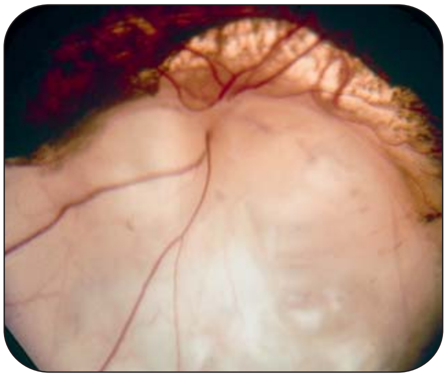

Fig. 9.27

Acquired

Retinoblastoma

Definition

- Retinoblastoma is the most common primary intraocular malignancy of childhood. In hereditary cases the tumour is bilateral and often multifocal; non-hereditary tumours are unilateral.

Presentation

- Within the first year of life in bilateral cases and around 2 years in unilateral (**Fig. 9.28**), with an average of 18 months.

Signs

- Endophytic tumours grow into the vitreous cavity (**Fig. 9.29**).
- Exophytic tumours grow in the subretinal space and cause retinal detachment (**Fig. 9.30**).

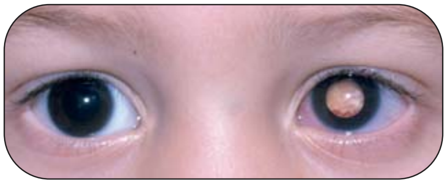
Fig. 9.28

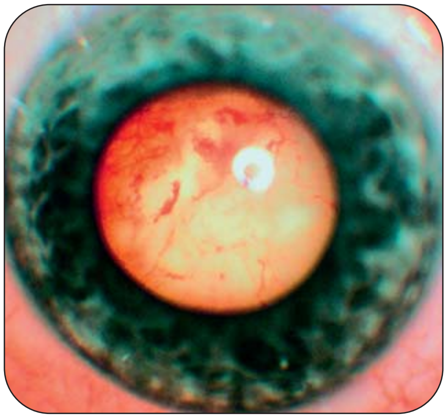
Fig. 9.29

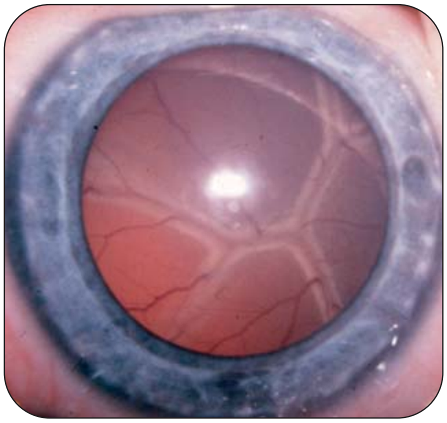
Fig. 9.30

Cicatricial retinopathy of prematurity

Presentation

- Retinopathy of prematurity is now a rare cause of leukocoria that presents at about 4 months.

Signs

- Retrolental fibrovascular tissue and retinal detachment (retrolental fibroplasia – **Fig. 9.31**) that may be bilateral.

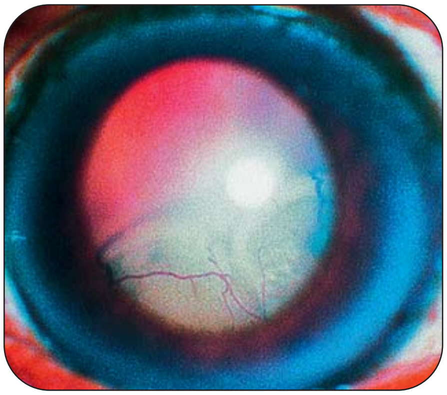
Fig. 9.31

Chronic toxocaral endophthalmitis

Presentation

- Between the ages of 2 and 9 years and is always unilateral (**Fig. 9.32**).

Signs

- Anterior uveitis and posterior synechiae (**Fig. 9.33**).
- Cyclitic membrane and low intraocular pressure.
- Vitreous opacification and peripheral retinal exudation.

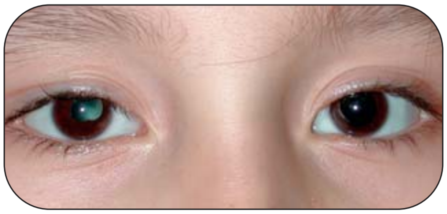

Fig. 9.32

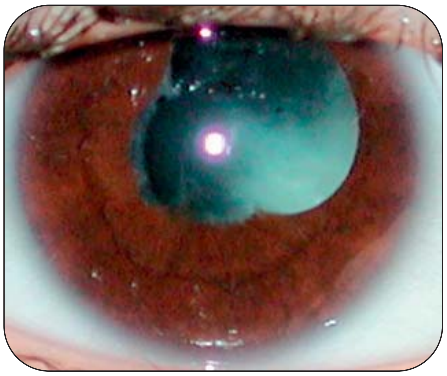

Fig. 9.33

Coats disease

Definition

- An idiopathic unilateral retinal telangiectasia (**Fig. 9.34**) that typically affects boys.

Presentation

- In the 1st decade (average 5 years).

Signs

- Exudative retinal detachment and surface telangiectasia.
- Massive subretinal lipid exudation (**Fig. 9.35**).

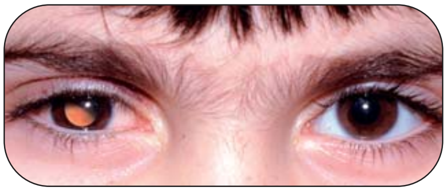

Fig. 9.34

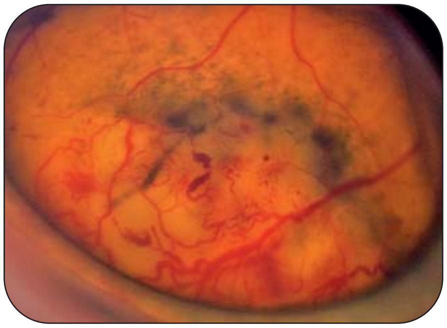

Fig. 9.35

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Lens 10

ABNORMALITIES OF SHAPE 213
Coloboma 213
Microphakia 213
Microspherophakia 213
Lentiglobus 214
Anterior lenticonus 214
Posterior lenticonus 214
ECTOPIA LENTIS 215
Developmental 215
Ocular associations 215
Hereditary causes without systemic manifestations 215
Marfan syndrome 215
Weill–Marchesani syndrome 216
Homocystinuria 216
Other causes 217
Acquired 217
CHILDHOOD CATARACT 218
Morphology 218
Anterior polar 218
Posterior polar 218
Coronary (supranuclear) 219
Lamellar 219
Central pulverulent 220
Sutural 220
Focal blue dot 220
Causes and associations of cataracts in neonates 221
Idiopathic 221
Hereditary 221
Associated with other ocular anomalies 221
Lowe oculocerebrorenal syndrome 221
Intrauterine infections 222
Galactosaemia 222
Hallermann–Streiff–François syndrome 223
Other causes 223
Causes and associations of cataract in older children 224
Idiopathic and hereditary 224
Galactokinase deficiency 224
Down syndrome (trisomy 21) 224
Fabry disease 224
Mannosidosis 225
ACQUIRED CATARACT 225
Anterior capsular 225
True exfoliation 225
Pseudoexfoliation syndrome 225
Pigmentation on anterior capsule 226
Anterior subcapsular 226
Glaukomflecken 226
Fibrous plaques 226
Posterior subcapsular 227
Chronic intraocular inflammation 227
Posterior segment disease 227
Drug-induced 227
Electric shock 227
Radiation 227
Neurofibromatosis 2 228
Cortical 228
Cuneiform 228
Snowflake 228
Christmas tree 229
Stellate 229
Flower-shaped 229
Nuclear 230
NON-OPAQUE LENS DEFECTS 230
Retro-dots 230
Water clefts 231
Vacuoles 231
OPACITIES IN PSEUDOPHAKIA 231
Capsular opacification 231
Posterior capsular fibrosis 231
Elschnig pearls 232
Anterior capsular fibrosis and contraction (phimosis) 232
Soemmering ring 232
Inflammatory plaques 232
Pseudo-lenticular opacities 233
Inflammatory precipitates 233
Pitting 233
Total opacification 234

ABNORMALITIES OF SHAPE

Coloboma

Definition

- A rare, congenital, unilateral or bilateral, condition which may be associated with other defects.

Signs

- Notching of the inferior or inferotemporal equator with absent zonules (**Fig. 10.1**).

Look for

- Other colobomas and microphthalmos (**Fig. 10.2**).
- Ciliary body medulloepithelioma which may cause acquired notching of the lens.

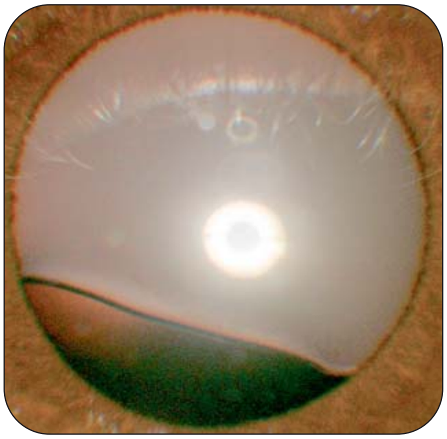

Fig. 10.1

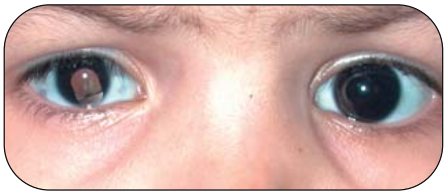

Fig. 10.2

Microphakia

Definition

- A small lens (**Fig. 10.3**) that occurs in association with conditions that cause arrest of ocular development such as microphthalmos.
- It may, however, be an isolated finding or associated with Lowe syndrome (see below).

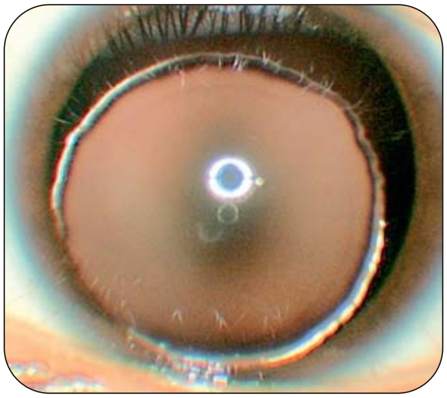

Fig. 10.3

Microspherophakia

Definition

- A rare, congenital, bilateral, condition that may be associated with a variety of other disorders (Table 10.1).

Signs

- The lens has a small diameter and spherical shape, resulting in lenticular myopia (**Fig. 10.4**).
- Ectopia is usually downwards or into the anterior chamber.

Table 10.1 **Associations of microspherophakia**

1. **Systemic syndromes**
   - Weill–Marchesani
   - Marfan
2. **Miscellaneous**
   - AD familial
   - Hyperlysinaemia
   - Familial ectopia lentis et pupillae
   - Peters anomaly
   - Congenital rubella

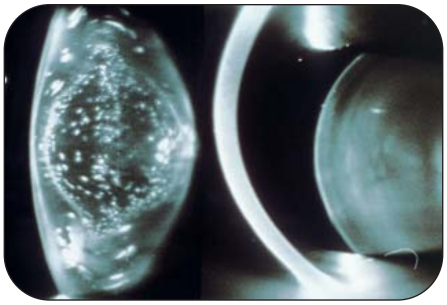

Fig. 10.4

Lentiglobus

Definition

- A very rare, congenital, usually unilateral, condition which causes induced myopia.

Signs

- A generalised hemispheric lens deformity which may be associated with posterior polar lens opacity.

Anterior lenticonus

Definition

- A rare, congenital, bilateral, condition which is associated with Alport syndrome which may be AD or X-LR.

Signs

- Localised anterior central conical projection (**Fig. 10.5**) that may later be associated with opacification.
- In early cases the red reflex shows an 'oil-droplet' sign (**Fig. 10.6**) similar to that seen in keratoconus.

Look for

- Posterior polymorphous corneal dystrophy.
- Flecked retina (see Figs 13.232 and 13.233).
- Progressive deafness and renal disease.

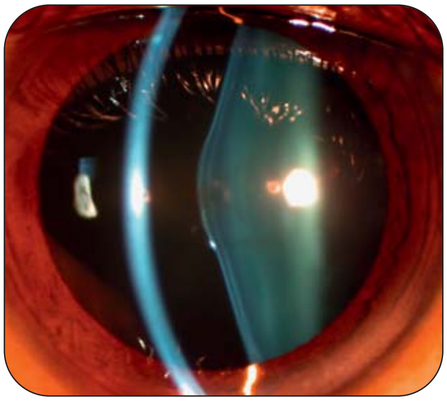

Fig. 10.5

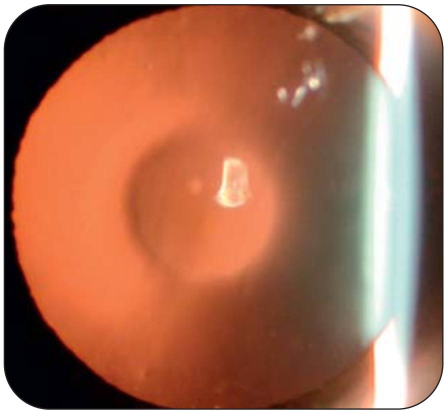

Fig. 10.6

Posterior lenticonus

Definition

- A rare, congenital, bilateral, condition which may occur in isolation or in association with Lowe syndrome.

Signs

- Localised posterior conical projection of the lens (**Fig. 10.7**).
- Associated posterior with opacification is common (**Fig. 10.8**).

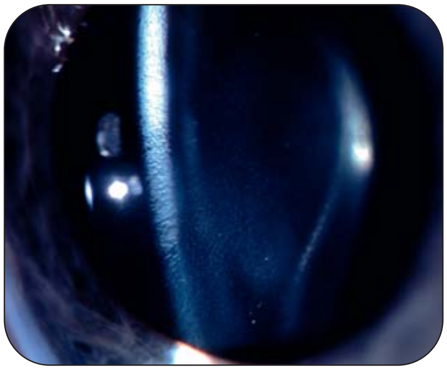

Fig. 10.7

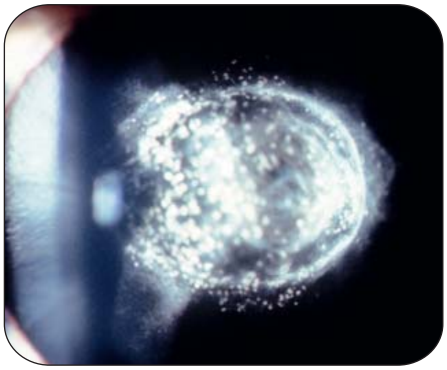

Fig. 10.8

Look for

- Psychomotor retardation.
- Muscular hypotonia.
- Renal disease.

ECTOPIA LENTIS

Developmental

Ocular associations

a. *Megalocornea.*

b. *Severe buphthalmos.*

c. *Very high myopia.*

d. *Aniridia* (**Fig. 10.9**).

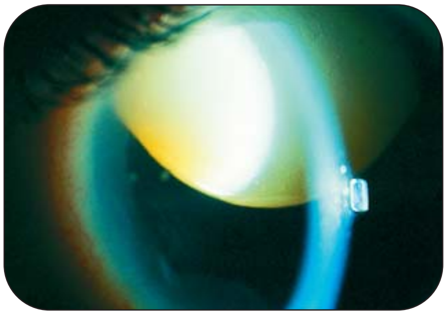

Fig. 10.9

Hereditary causes without systemic manifestations

a. *AD familial ectopia lentis.*

b. *AR ectopia lentis et pupillae* in which pupil and lens are displaced in opposite directions (see Fig. 9.16).

c. *Isolated familial microspherophakia.*

Marfan syndrome

Definition

- An uncommon AD widespread disorder of connective tissue.

Signs

- Bilateral, symmetric, slowly progressive usually superotemporal subluxation with intact zonule (**Fig. 10.10**) usually noted in childhood or early adult life.
- Total dislocation into the vitreous (**Fig. 10.11**) may occasionally occur in older patients.

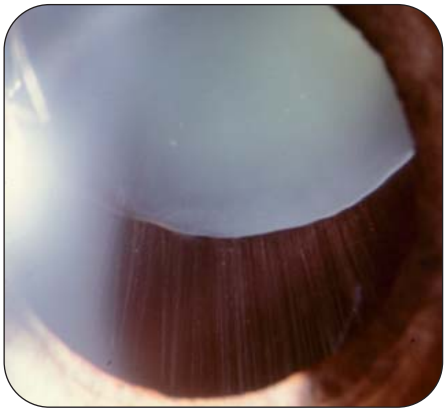

Fig. 10.10

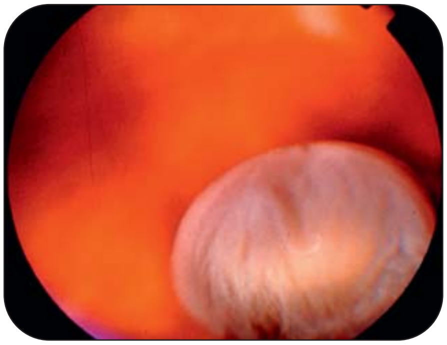

Fig. 10.11

Look for

- Retinal detachment.
- Angle anomaly and glaucoma.
- Tall and thin stature with arm span > height.
- Arachnodactyly (**Fig. 10.12**).
- High-arched palate (see Fig. 15.38).
- Cardiovascular disease.

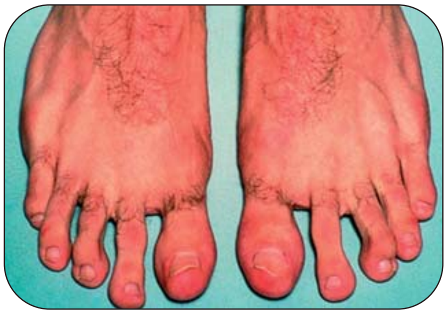

Fig. 10.12

Weill–Marchesani syndrome

Definition

- A rare, usually AR, disease of connective tissue, conceptually the reverse of Marfan syndrome.

Signs

- Bilateral inferior subluxation may occur in late childhood or early adult life.
- Subluxation into the anterior chamber may also occur because the lens is small (**Fig. 10.13**).

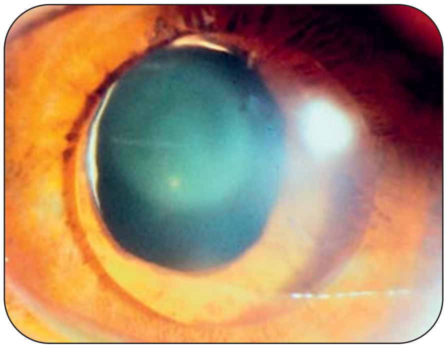

Fig. 10.13

Look for

- Short stature.
- Small, stubby toes (**Fig. 10.14**) and fingers (brachydactyly).
- Stiff joints.
- Mental handicap.

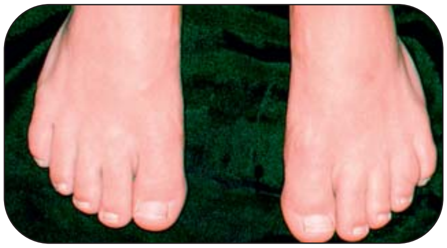

Fig. 10.14

Homocystinuria

Definition

- A rare AR condition caused by an enzyme defect.

Signs

- Typically inferonasal subluxation with disintegration of the zonules (**Fig. 10.15**) occurs in all patients by the age of 25 years.
- Lens incarceration in the pupil (see Fig. 7.14) or total subluxation into the anterior chamber may also occur.

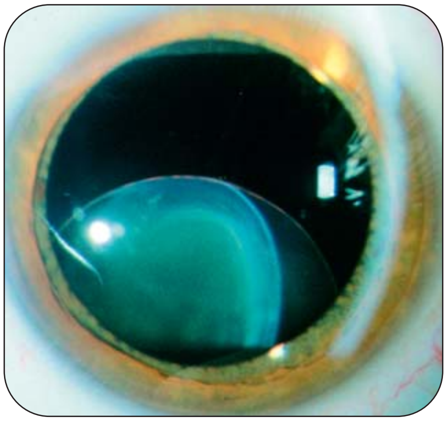

Fig. 10.15

Look for

- Coarse fair hair and a malar flush (**Fig. 10.16**).
- Marfanoid habitus.
- Tendency to thromboses.

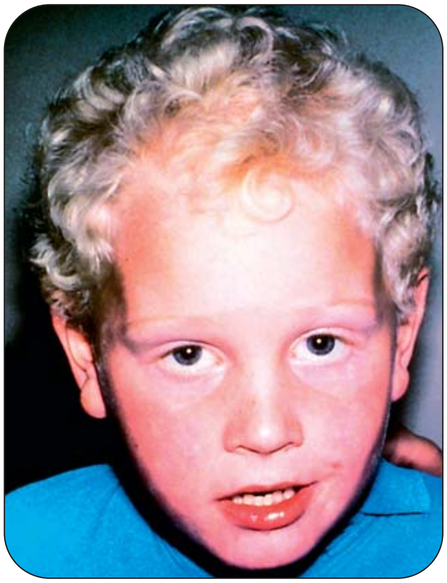

Fig. 10.16

Other causes

a. *Ehlers–Danlos syndrome type 6* (ocular sclerotic) is an AR connective tissue disorder in which ectopia lentis is uncommon.

b. *Hyperlysinaemia* is an AR inborn error of metabolism which is occasionally associated with ectopia lentis.

c. *Stickler syndrome* is an AD condition in which ectopia lentis is uncommon.

d. *Sulfite oxidase deficiency* is an AR disorder of sulphur metabolism in which ectopia lentis is universal.

Acquired

a. *Blunt ocular trauma* may cause subluxation and rarely dislocation into the anterior chamber.

b. *Ciliary body melanoma* may cause a slight shift of the lens as well as a localised opacity (**Fig. 10.17**).

c. *Hypermaturity* in which the cortex has liquefied and the hard nucleus sinks inferiorly (**Fig. 10.18**); less frequently, the entire lens may be subluxated (**Fig. 10.19**).

d. *Degenerated eye* (**Fig. 10.20**).

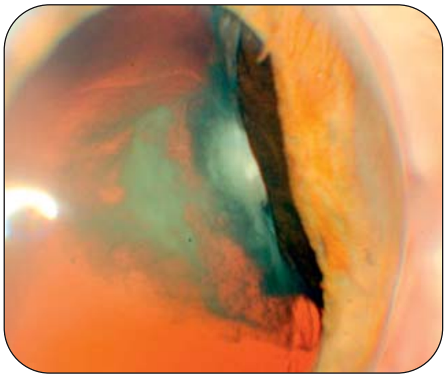

Fig. 10.17

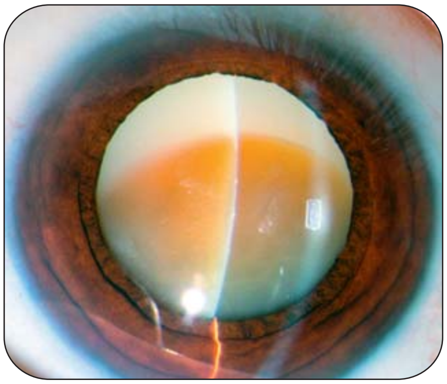

Fig. 10.18

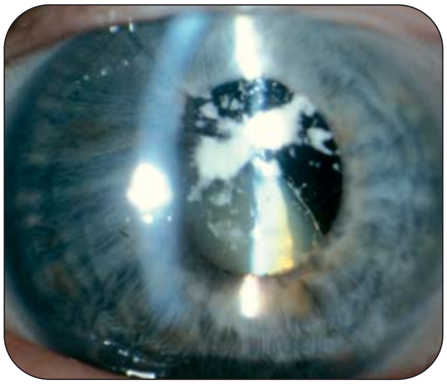

Fig. 10.19

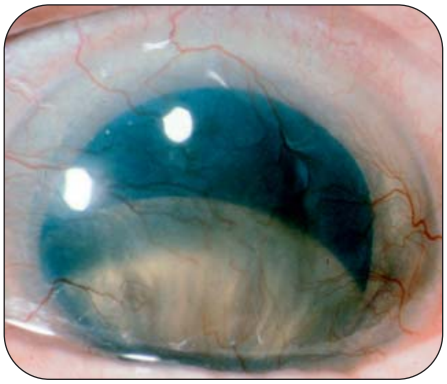

Fig. 10.20

CHILDHOOD CATARACT

Morphology

Anterior polar

Inheritance

- Usually AD.

Signs

- The opacity may involve only the capsule (**Fig. 10.21**).
- It may be pyramidal and project into the anterior chamber (**Fig. 10.22**).

Look for

- Persistent pupillary membrane (**Fig. 10.23**).
- Anterior lenticonus.
- Peters anomaly.
- Microphthalmos.
- Aniridia.

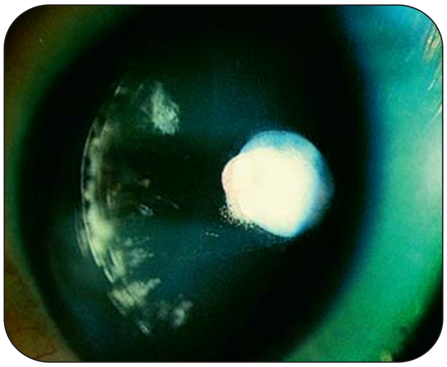
Fig. 10.21

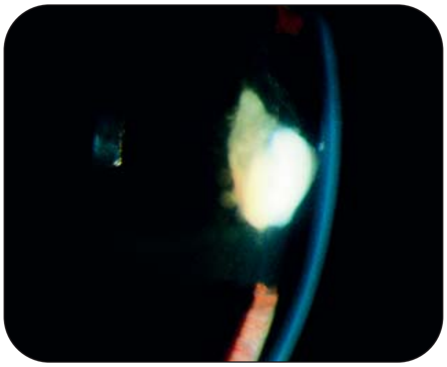
Fig. 10.22

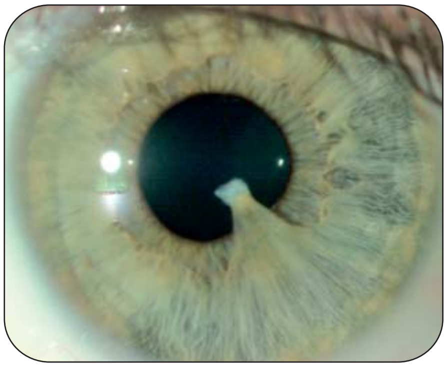
Fig. 10.23

Posterior polar

Signs

- The opacity that may involve only the capsule or it may form a plaque.

Look for

- Persistent hyaloid remnants (Mittendorf dot – **Fig. 10.24**).
- Posterior lenticonus (see Fig. 10.8).
- Persistent anterior fetal vasculature.

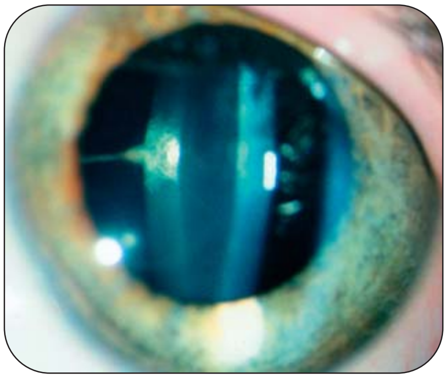
Fig. 10.24

Coronary (supranuclear)

Signs

- A deep cortical opacity that surrounds the nucleus like a crown (**Fig. 10.25**).

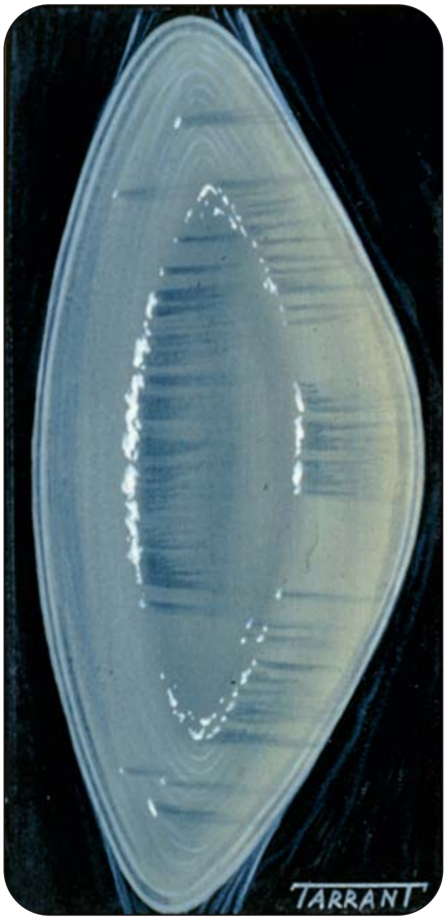

Fig. 10.25

Lamellar

Inheritance

- Usually AD, occasionally AR.

Signs

- Round, central, shell-like opacity surrounding the nucleus (**Fig. 10.26**).
- The opacity may be associated with riders and may progress to involve the entire nucleus (**Fig. 10.27**).

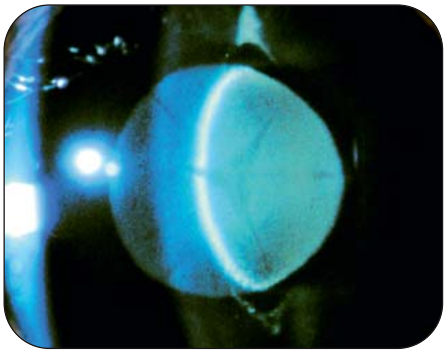

Fig. 10.26

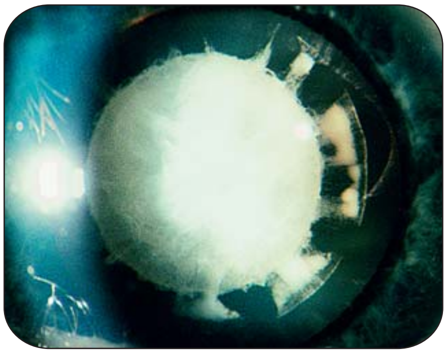

Fig. 10.27

Look for

- Galactosaemia.
- Hypoglycaemia.
- Hypocalcaemia.

Central pulverulent

Inheritance

- AD.

Signs

- Spheroidal nuclear opacity with a clearer centre (**Fig. 10.28**).

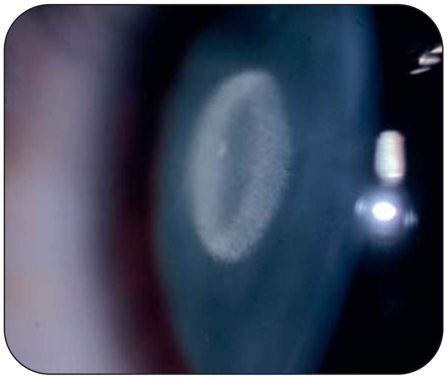

Fig. 10.28

Sutural

Inheritance

- XL.

Signs

- The opacity follows the shape of the anterior or posterior Y suture (**Fig. 10.29**).

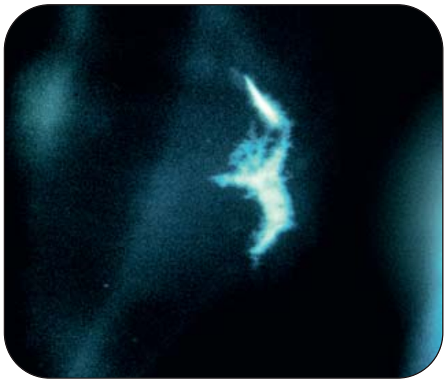

Fig. 10.29

Focal blue dot

Signs

- Common and innocuous opacities that may coexist with other types of congenital cataracts (**Fig. 10.30**).

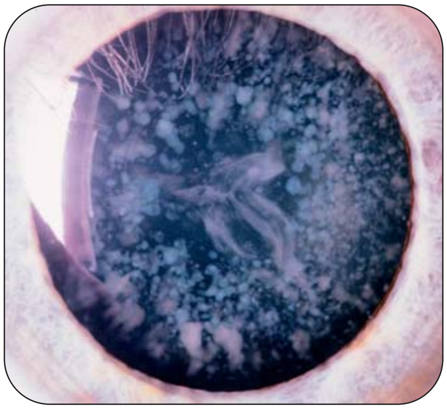

Fig. 10.30

Causes and associations of cataracts in neonates

Idiopathic

Approximately 50% of bilateral infantile cataract cases are idiopathic and 90% of unilateral cataracts are idiopathic.

Hereditary

a. *Anterior polar* – AD.

b. *Lamellar* – usually AD, occasionally AR.

c. *Central pulverulent* – AD.

d. *Sutural* – XL.

Look for

- Similar opacities in family members.

Associated with other ocular anomalies

a. *Aniridia* (see Fig. 10.9).

b. *Coloboma* (**Fig. 10.31**).

c. *Microphthalmos* and *persistent pupillary membrane* (**Fig. 10.32**).

d. *Peters anomaly* (**Fig. 10.33**).

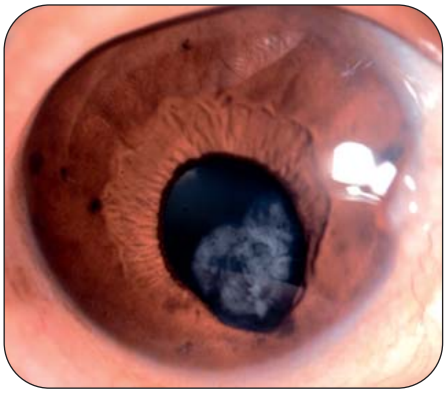

Fig. 10.31

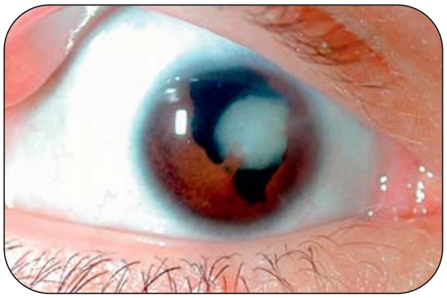

Fig. 10.32

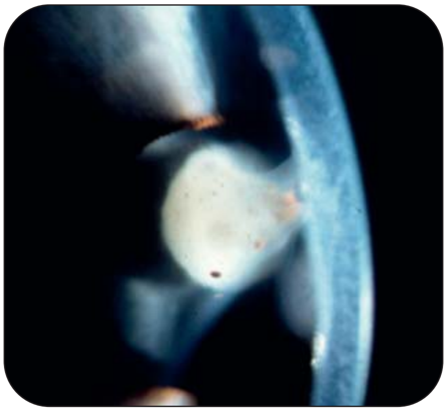

Fig. 10.33

Lowe oculocerebrorenal syndrome

Definition

- A very rare XL inborn error of amino acid metabolism.
- It is associated with microphakia and a variety of lens opacities, as well as glaucoma in 50% of cases.

Look for

- Renal disease.
- Muscular hypotonia.
- Frontal prominence and sunken eyes (**Fig. 10.34**).

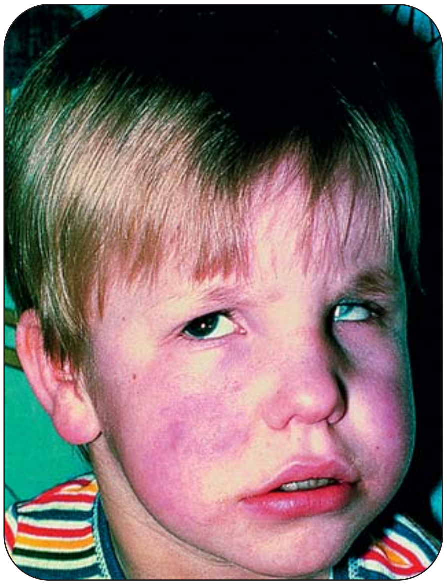

Fig. 10.34

Intrauterine infections

Signs

- Dense, central, unilateral (**Fig. 10.35**) or bilateral opacities.

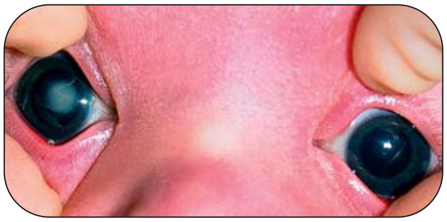

Fig. 10.35

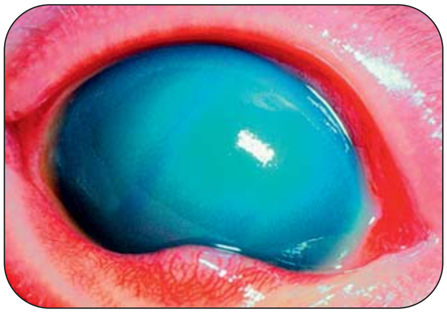

Fig. 10.36

Look for

- Microphthalmos.
- Corneal opacification and glaucoma (**Fig. 10.36**).
- History of maternal illness in pregnancy associated with a rash which is suggestive of intrauterine rubella or varicella.

Galactosaemia

Definition

- A rare AR inborn error of metabolism caused by an enzyme defect.

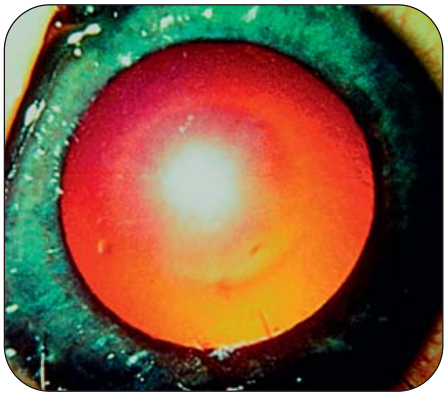

Fig. 10.37

Signs

- The lens has a characteristic central oil-droplet appearance (**Fig. 10.37**).
- Unless treated with a galactose-restricted diet, lamellar opacities develop and maturity may ensue.

Look for

- Failure to thrive and lethargy (**Fig. 10.38**).
- Vomiting and diarrhoea.
- Reducing substance in the urine.

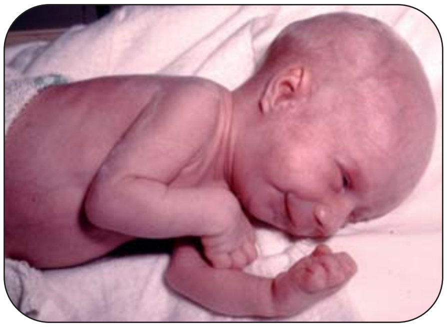

Fig. 10.38

Hallermann–Streiff–François syndrome

Definition

- A rare sporadic disorder associated with cataract in the vast majority of patients.

Signs

- The cataract is often membranous, the lens material having been resorbed leaving residual chalky lens material sandwiched between the anterior and posterior capsule (**Fig. 10.39**).

Look for

- Frontal prominence, sparse hair, micrognathia and beaked nose (**Fig. 10.40**).
- Progeria, short stature and hypodontia.

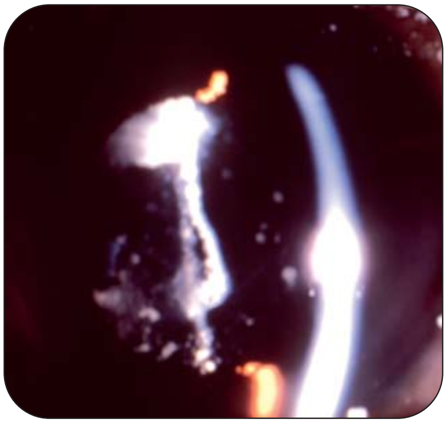

Fig. 10.39

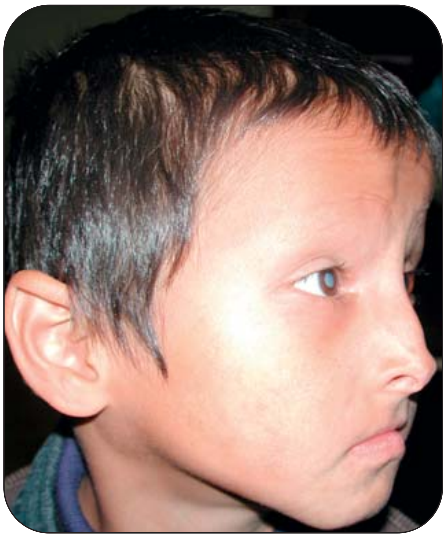

Fig. 10.40

Other causes

a. *Maternal radiation* during the first trimester of pregnancy.

b. *Neonatal hypoglycaemia.*

c. *Neonatal hypocalcaemia*.

Causes and associations of cataract in older children

Idiopathic and hereditary (see above)

Galactokinase deficiency

- Cataract may be the only manifestation.

Down syndrome (trisomy 21)

Signs

- A variety of lens opacities that usually develop in late childhood.

Look for

- Brushfield spots (see Fig. 8.40).
- Typical facies (**Fig. 10.41**).
- Broad stubby hands with a single (simian) crease.
- Mental handicap.
- Cardiac malformation.

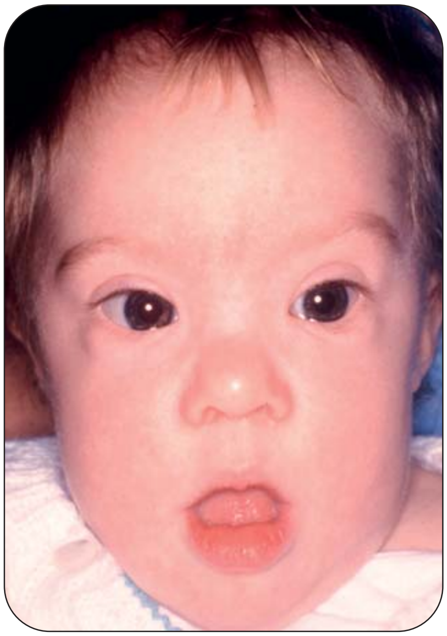

Fig. 10.41

Fabry disease

Inheritance

- XL-R.

Signs

- Cortical spoke-like opacities.

Look for

- Vortex keratopathy.
- Retinal venous tortuosity.
- Third nerve palsy and nystagmus.
- Periodic painful extremities.
- Purple cutaneous telangiectasia (angiokeratoma diffusum – **Fig. 10.42**).
- Renal and cardiac disease.
- Periodic pain in extremities.
- Renal and cardiovascular disease.

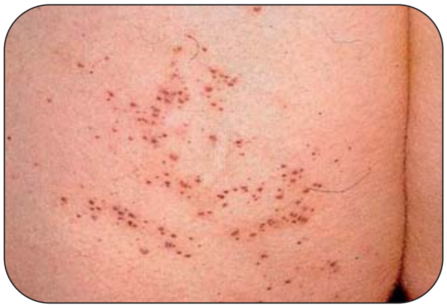

Fig. 10.42

Mannosidosis

Definition

- A very rare AR disease caused by an enzyme deficiency.

Signs

- Cortical spoke-like opacities.

Look for

- Corneal clouding.
- Psychomotor retardation.
- Susceptibility to infection.
- Deafness.

Table 10.2 **Systemic associations of childhood cataract**

1. **Maternal intrauterine infections**
    - Rubella
    - Toxoplasmosis
    - Cytomegalovirus
    - Varicella
2. **Metabolic disorders**
    - Galactosaemia
    - Galactokinase deficiency
    - Hypoglycaemia
    - Hypocalcaemia
    - Lowe syndrome
    - Congenital haemolytic jaundice
3. **Central nervous system abnormalities**
    - Zellweger syndrome
    - Sjögren–Larsson syndrome
    - Norrie disease
4. **Craniofacial syndromes**
    - Crouzon
    - Apert
    - Pierre Robin
    - Hallermann–Streiff–François
    - Stickler
5. **Chromosomal abnormalities**
    - Down syndrome (trisomy 21)
    - Patau syndrome (trisomy 13)
    - Edward syndrome (trisomy 18)
    - Cri-du-chat syndrome (deletion of short arm of chromosome 5)
6. **Skin abnormalities**
    - Cockayne syndrome
    - Congenital ichthyosis

ACQUIRED CATARACT

Anterior capsular

True exfoliation

Cause

- This is now an extremely rare finding caused by exposure to infrared radiation (glass-blowers, furnace workers and desert dwellers).

Signs

- Lamellar splits in the anterior capsule forming a scroll-like membrane (**Fig. 10.43**).

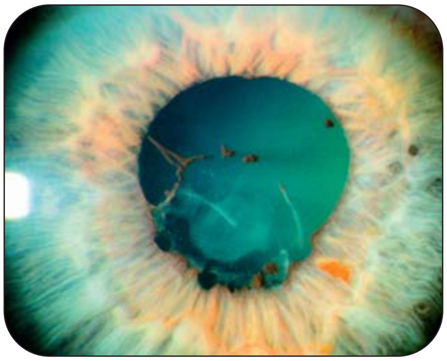

Fig. 10.43

Pseudoexfoliation syndrome

Signs

- White material on the anterior lens capsule has a bull's-eye configuration (**Fig. 10.44**).

Look for

- Trabecular hyperpigmentation (see Fig. 7.1).
- Glaucoma.

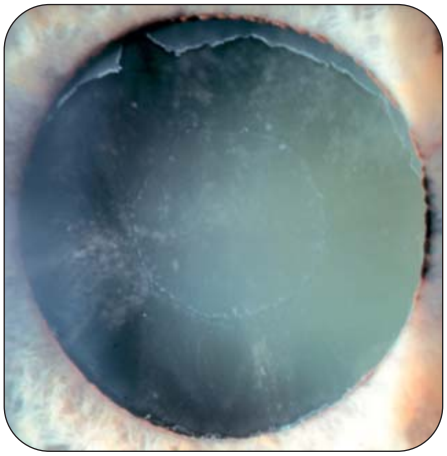

Fig. 10.44

Pigmentation on anterior capsule

a. *Following iritis* (**Fig. 10.45**).

b. *Blunt trauma* (Vossius ring – **Fig. 10.46**).

c. *Drug-induced* – chlorpromazine (**Fig. 10.47**), amiodarone, gold and tamoxifen.

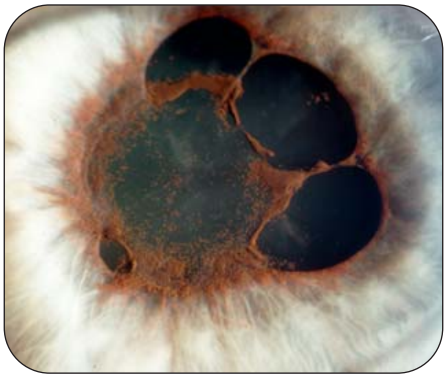
Fig. 10.45

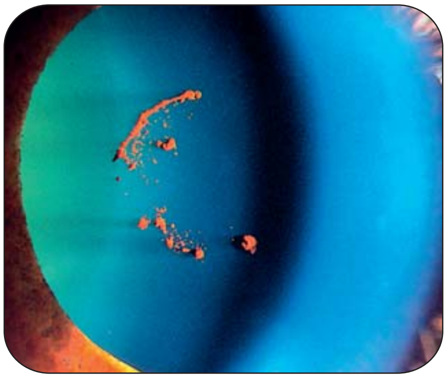
Fig. 10.46

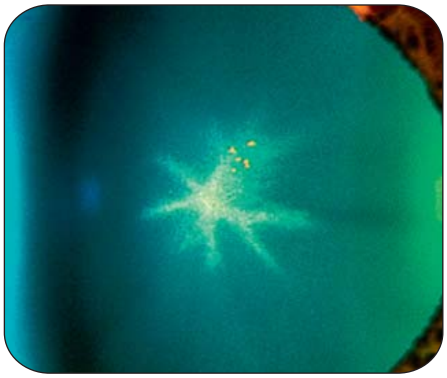
Fig. 10.47

Anterior subcapsular

Glaukomflecken

Cause

- Acute congestive attack of angle-closure glaucoma.

Signs

- Grey-white opacities in the pupillary zone which resemble ‘spilled milk’ (**Fig. 10.48**).

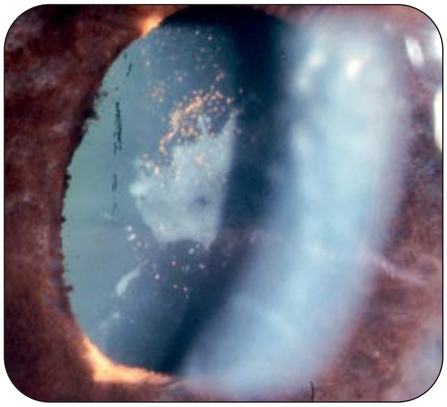
Fig. 10.48

Fibrous plaques

Causes

- Atopic dermatitis (**Fig. 10.49**).
- Chronic anterior uveitis (**Fig. 10.50**).

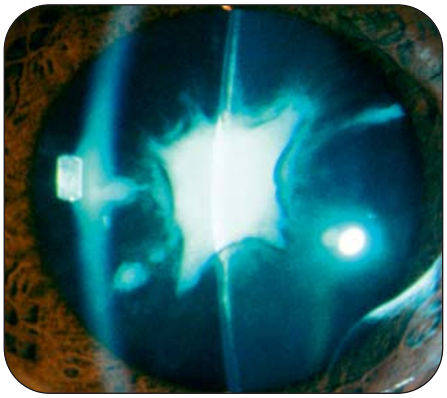
Fig. 10.49

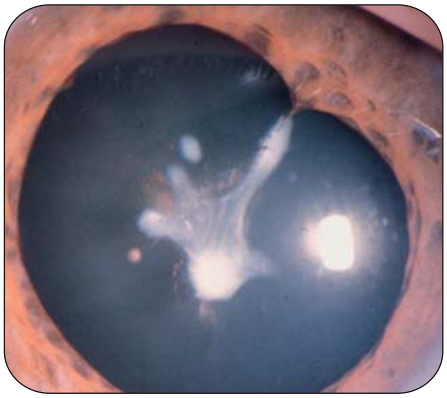
Fig. 10.50

Posterior subcapsular

Posterior subcapsular cataracts may be age-related or associated with the following conditions.

Chronic intraocular inflammation

Signs

- Initially polychromatic lustre of the posterior capsule (**Fig. 10.51**).
- Later subcapsular cataract (**Fig. 10.52**).

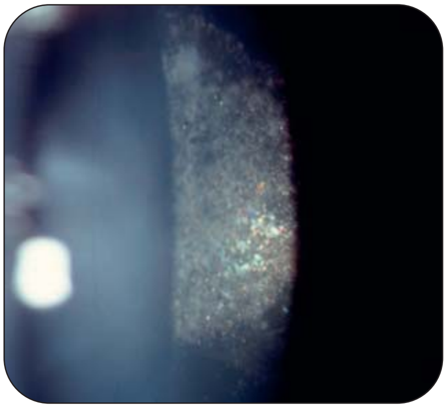

Fig. 10.51

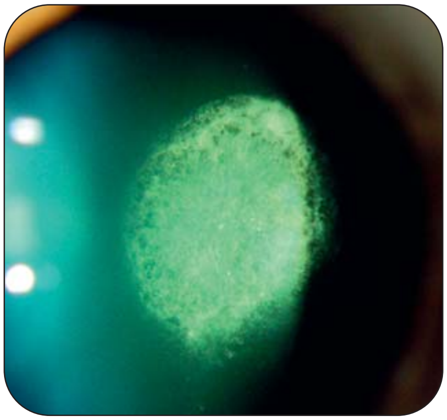

Fig. 10.52

Posterior segment disease

a. *Degenerative myopia*, which is also associated with nuclear cataract.

b. *Hereditary fundus dystrophies* such as retinitis pigmentosa and gyrate atrophy.

Drug-induced

a. *Topical and systemic steroids.*

b. *Systemic busulfan.*

Electric shock

Signs

- Delicate stellate-like subcapsular opacities (**Fig. 10.53**).

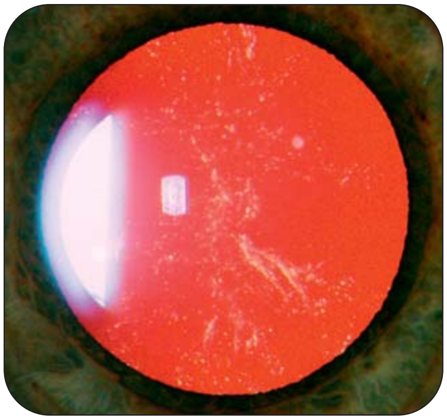

Fig. 10.53

Radiation

Radiation for ocular malignancies may cause lens opacities months or years later.

Neurofibromatosis 2

Definition

- An AD phacomatosis that is associated with cataract formation in 60% of patients prior to the age of 30 years.

Look for

- Bilateral acoustic neuromas (**Fig. 10.54**).

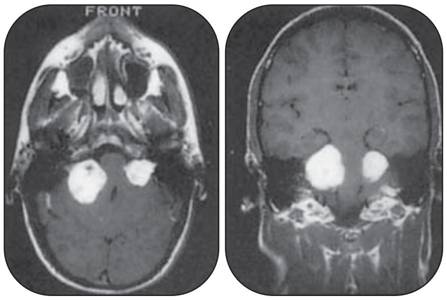

Fig. 10.54

Cortical

Cuneiform

Signs

- Spoke-like opacity best seen on retroillumination (**Fig. 10.55**) is the most common type of age-related cataract.

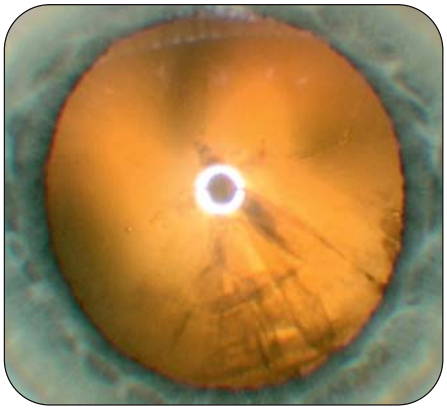

Fig. 10.55

Snowflake

Signs

- Typically affects young diabetic patients.
- Abrupt onset of minute cortical snowflake-like opacities (**Fig. 10.56**).
- Maturity may ensue within a few days (**Fig. 10.57**).

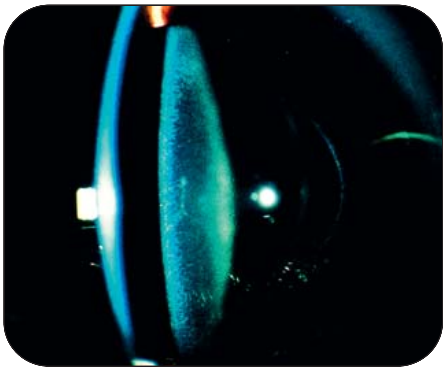

Fig. 10.56

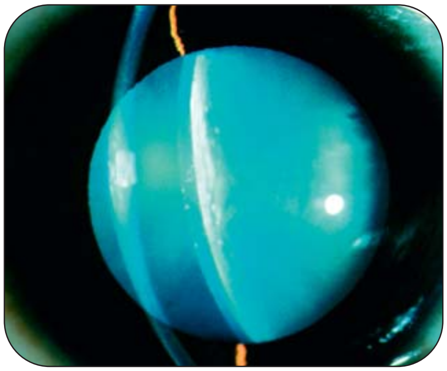

Fig. 10.57

Christmas tree

Signs

- Polychromatic, needle-like deposits in the deep cortex and nucleus (**Fig. 10.58**) is an uncommon age-related opacity.

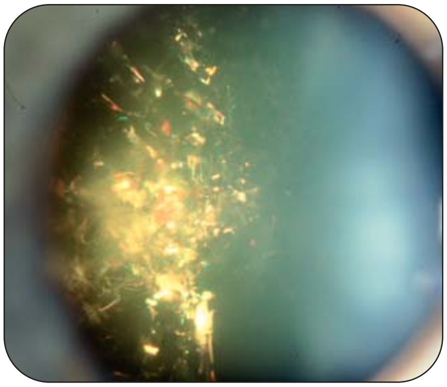

Fig. 10.58

Stellate

Signs

- Stellate posterior cortical cataract (**Fig. 10.59**) occurs in patients with myotonic dystrophy.

Look for

- Mournful facial expression and frontal balding (**Fig. 10.60** – note left cataract).
- Myotonia of peripheral muscles.
- Hypogonadism.

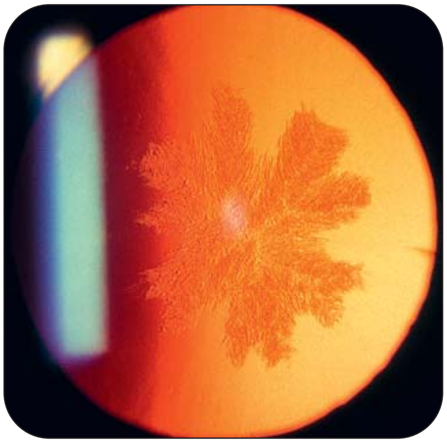

Fig. 10.59

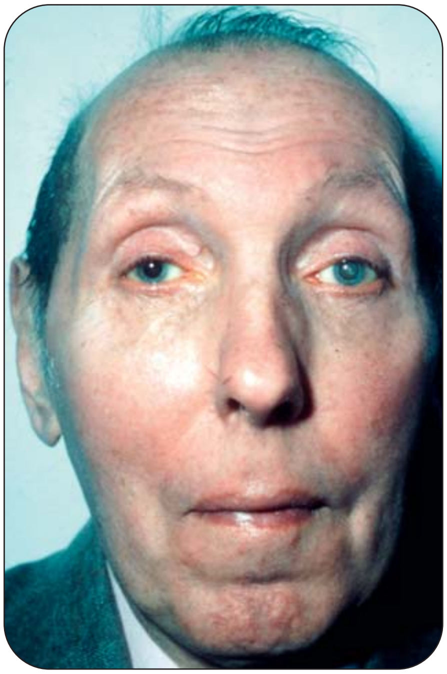

Fig. 10.60

Flower-shaped

Signs

- An unusual opacity (**Fig. 10.61**) caused by blunt trauma.

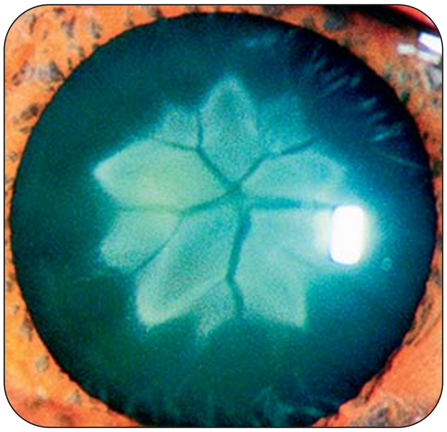

Fig. 10.61

Nuclear

Signs

- Nuclear cataracts vary in density and colour, from yellow (**Fig. 10.62**) to brown (**Fig. 10.63**), and very occasionally virtually black (cataract nigrans).
- The red reflex may be virtually normal even in moderately advanced cases (**Fig. 10.64**).

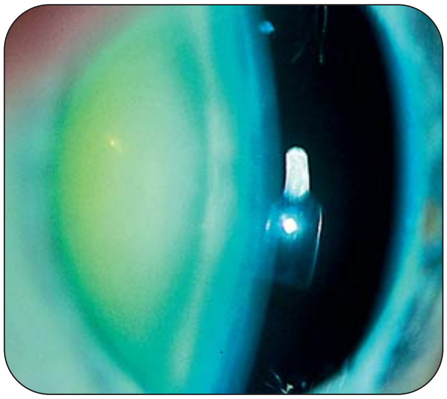
Fig. 10.62

Causes

- Age-related.
- Degenerative myopia.
- Post-vitrectomy.
- Diabetes may cause early-onset nuclear cataract.

Table 10.3 **Systemic associations of acquired cataract**

- Diabetes mellitus
- Atopic dermatitis
- Myotonic dystrophy
- Hypocalcaemia
- Wilson disease
- NF2

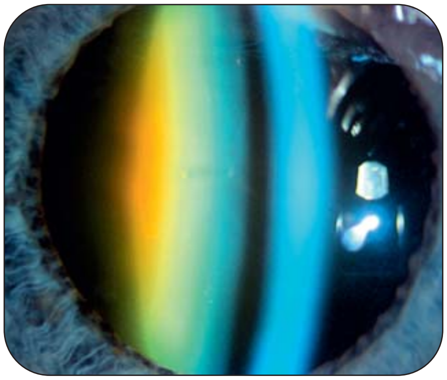
Fig. 10.63

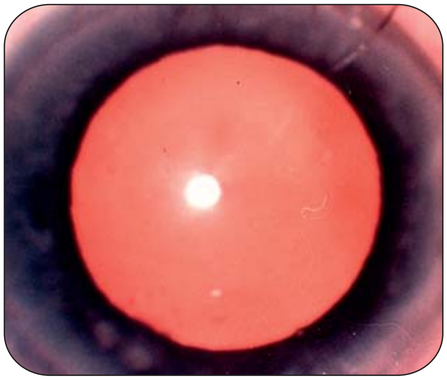
Fig. 10.64

NON-OPAQUE LENS DEFECTS

Non-opaque lens defects occur in association with cataract and may represent an aspect of the cataract forming process.

Retro-dots

Signs

- Small dots best seen on retroillumination (**Fig. 10.65**).

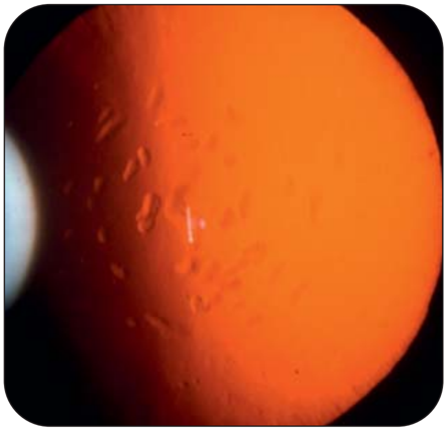
Fig. 10.65

Water clefts

Signs

- Radial, wedge-like defects in the cortex (**Fig. 10.66**) which may cause ghosting of images.

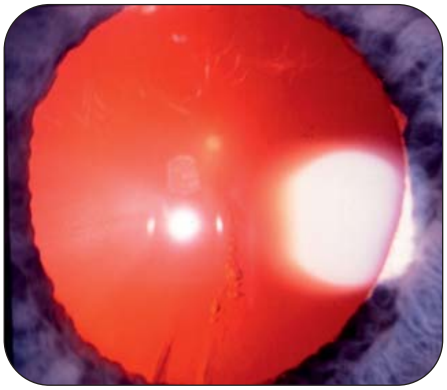

Fig. 10.66

Vacuoles

Signs

- Cystoid spaces in the superficial cortex (**Fig. 10.67**) that may coexist with other defects.

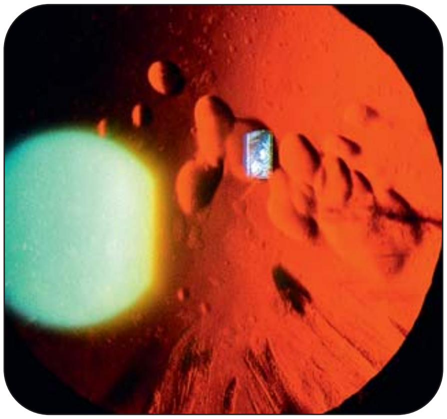

Fig. 10.67

OPACITIES IN PSEUDOPHAKIA

Capsular opacification

Posterior capsular fibrosis

Presentation

- Within 2–6 months of cataract surgery.

Signs

- Diffuse opacification and wrinkling of the posterior capsule (**Fig. 10.68**).

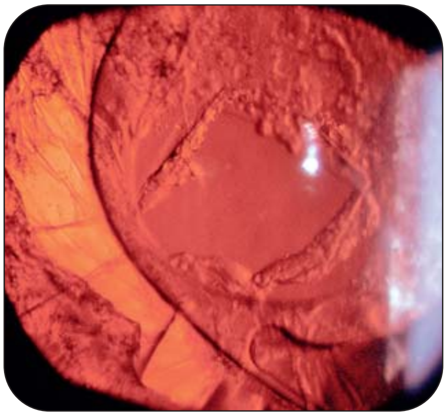

Fig. 10.68

Elschnig pearls

Cause

- Proliferation of lens epithelium onto the posterior capsule.

Presentation

- Several months or years after cataract surgery.

Signs

- Vacuolated appearance on the posterior capsule best seen on retroillumination (**Fig. 10.69**).

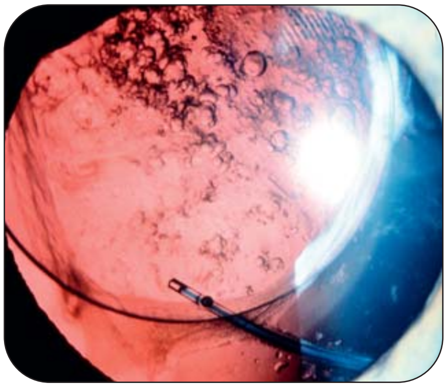

Fig. 10.69

Anterior capsular fibrosis and contraction (phimosis)

Presentation

- Several weeks after surgery.

Signs

- Severe opacification and radial wrinkling (**Fig. 10.70**).

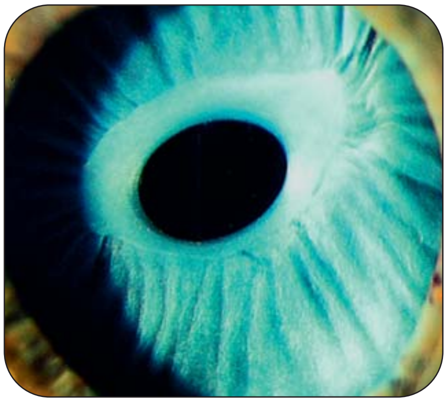

Fig. 10.70

Soemmering ring

Cause

- Sequestration of proliferating lens fibres in the equatorial region of the capsule.
- It occurred in the past following surgery for congenital cataract but is now rarely seen.

Sign

- Doughnut-shaped opacification (**Fig. 10.71**).

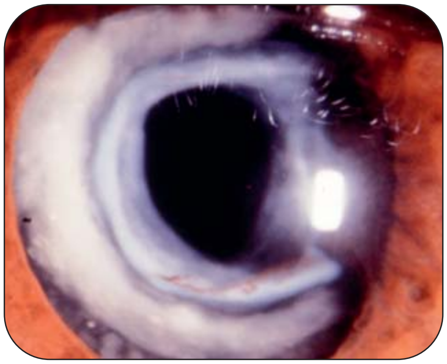

Fig. 10.71

Inflammatory plaques

Cause

- Chronic postoperative endophthalmitis.

Signs

- Localised fluffy white capsular deposits (**Fig. 10.72**).

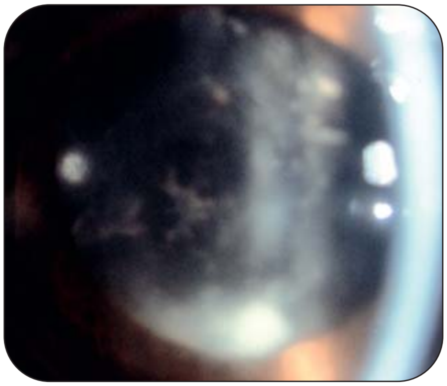

Fig. 10.72

Pseudo-lenticular opacities

Inflammatory precipitates

Signs

- Precipitates on the IOL (**Fig. 10.73**) usually associated with postoperative uveitis or endophthalmitis.

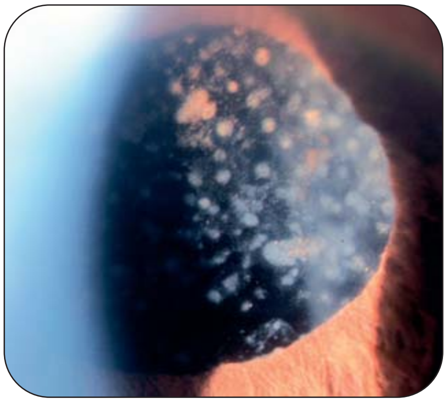

Fig. 10.73

Pitting

Signs

- Pitting of the IOL (**Fig. 10.74**) caused accidentally during YAG-laser posterior capsulotomy.

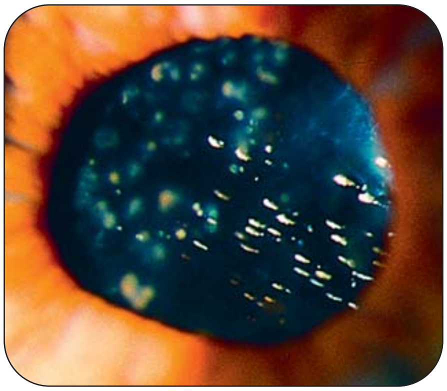

Fig. 10.74

Total opacification

Signs

- Total opacification (**Fig. 10.75**) of the IOL is very rare.

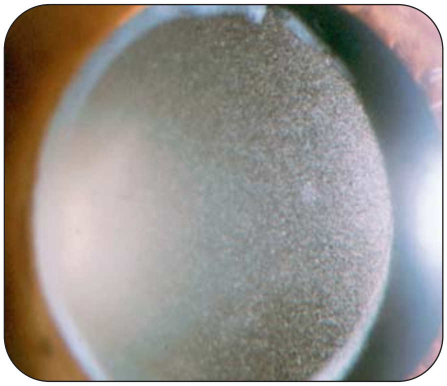

Fig. 10.75

Vitreous

11

VITREOUS CELLS **237**

Leucocytes **237**
Iridocyclitis *237*
Intermediate uveitis *237*
Fuchs syndrome *238*
Posterior and panuveitis *238*
Infectious endophthalmitis *238*

Erythrocytes **238**
Acute posterior vitreous detachment *238*
Proliferative retinopathies *239*
Miscellaneous retinal disorders *239*
Trauma *239*
Systemic causes *239*

Pigment cells **240**

Tumour cells **240**
Endophytic retinoblastoma *240*
Primary intraocular lymphoma *240*

VITREOUS OPACITIES **241**

Common floaters **241**
Innocuous floaters *241*
Associated with acute posterior vitreous detachment *241*

Inflammatory **241**
Cotton balls *241*
Other *242*

Iridescent particles **242**
Asteroid hyalosis (Benson disease) *242*
Synchysis scintillans *242*
Deposits in familial amyloidosis *243*

Miscellaneous **243**
Solitary retinal fragments (operculum) *243*
Congenital remnants *243*
Acquired cysts *244*
Lens fragments *244*
Foreign bodies and material *244*

HEREDITARY VITREORETINOPATHIES **245**

Associated with vitreous condensations **245**
Familial exudative vitreoretinopathy *245*
Autosomal dominant vitreoretinochoroidopathy *245*
Autosomal dominant neovascular inflammatory vitreoretinopathy *245*
Erosive vitreoretinopathy *245*

Associated with 'empty' vitreous **246**
Stickler syndrome *246*
Wagner disease *247*

VITREOUS CELLS

Leucocytes

Iridocyclitis

Signs

- Retrolental cells (**Fig. 11.1**) seen in association with aqueous cells.

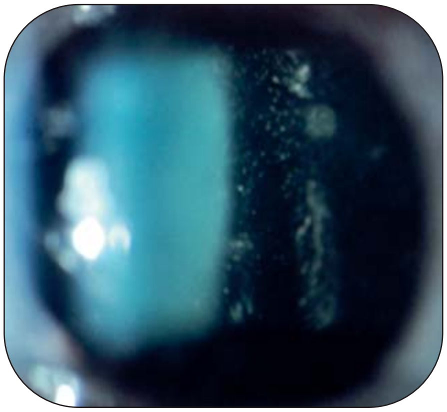

Fig. 11.1

Intermediate uveitis

Definition

- A common, chronic, frequently bilateral but often asymmetric condition that typically affects young adults. It may be idiopathic or associated with underlying systemic disease (Table 11.1).

Signs

- Vitreous cells (vitritis) vary in severity from mild to very severe (**Fig. 11.2**).

Look for

- Mild anterior uveitis.
- Inferior peripheral snowbanking (pars planitis – **Fig. 11.3**).
- Vitreous cotton balls.
- Peripheral periphlebitis.
- Chronic cystoid macular oedema.

Table 11.1 **Systemic associations of intermediate uveitis**

- Sarcoidosis
- Multiple sclerosis
- Whipple disease
- Lyme disease
- Cat-scratch fever
- Waldenström macroglobulinaemia

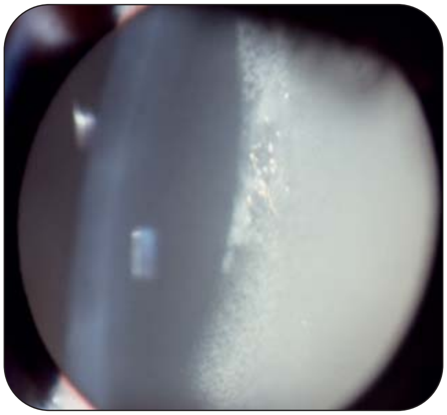

Fig. 11.2

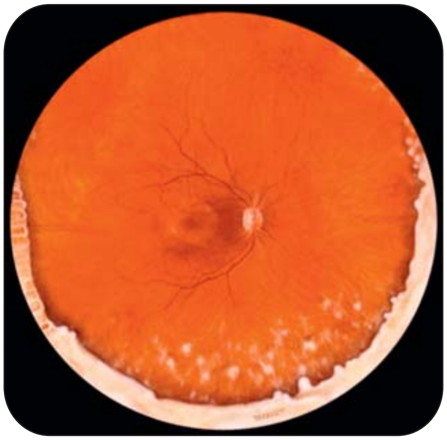

Fig. 11.3

Fuchs syndrome

Fuchs syndrome is frequently associated with vitritis that must not be mistaken for intermediate uveitis.

Posterior and panuveitis

Signs

- Cells are most dense at the site of the inflammatory focus (**Fig. 11.4**).
- Severe vitritis may impair visualisation of the fundus (**Fig. 11.5**).

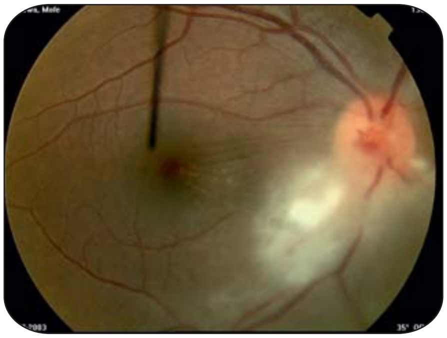

Fig. 11.4

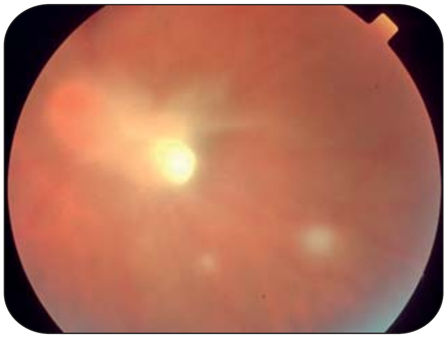

Fig. 11.5

Infectious endophthalmitis

Classification

a. *Acute-onset postsurgical and post-traumatic.*

b. *Delayed-onset chronic postoperative.*

c. *Delayed-onset acute associated with a filtering bleb.*

d. *Endogenous.*

Erythrocytes

Acute posterior vitreous detachment

Presentation

- Unilateral, sudden shower of minute red-coloured or dark spots frequently associated with photopsia.

Look for

- Source of bleeding is either a tractional retinal tear (**Fig. 11.6**) or less frequently avulsion of a peripheral blood vessel unassociated with a tear.

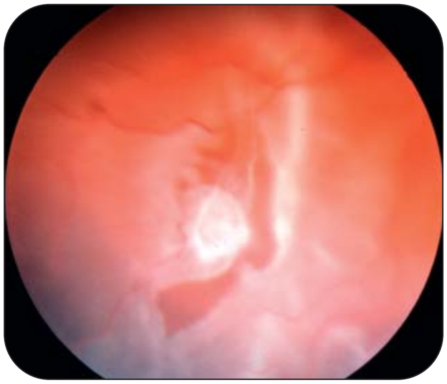

Fig. 11.6

Proliferative retinopathies

New vessel formation associated with the following conditions may cause vitreous haemorrhage.

a. *Diabetes* (**Fig. 11.7**).

b. *Following retinal vein occlusion.*

c. *Sickle cell disease.*

d. *Eales disease.*

e. *Vasculitis.*

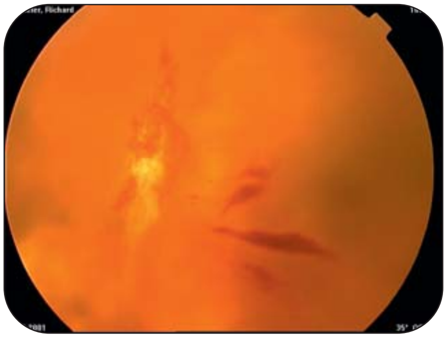

Fig. 11.7

Miscellaneous retinal disorders

a. *Macroaneurysm* (**Fig. 11.8**).

b. *Telangiectasia.*

c. *Capillary haemangioma.*

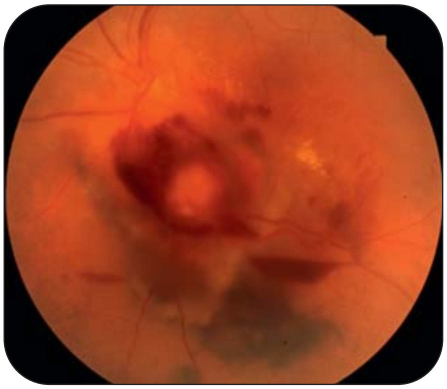

Fig. 11.8

Trauma

a. *Blunt* trauma which may be associated with dialysis formation (**Fig. 11.9**).

b. *Penetrating.*

c. *Iatrogenic* during surgery for retinal detachment.

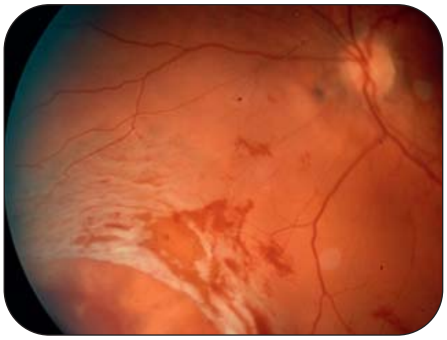

Fig. 11.9

Systemic causes

a. *Bleeding disorders.*

b. *Terson syndrome* (subarachnoid haemorrhage + vitreous haemorrhage).

Pigment cells

Signs

- Pigment cells ('tobacco dust') in the anterior vitreous (**Fig. 11.10**) consisting of macrophages containing retinal pigment epithelial cells.

Causes

- Acute retinal tear associated with acute posterior vitreous detachment (see Fig. 11.6).
- Rhegmatogenous retinal detachment.
- Excessive retinal cryotherapy.
- Proliferative vitreoretinopathy.
- Trauma.

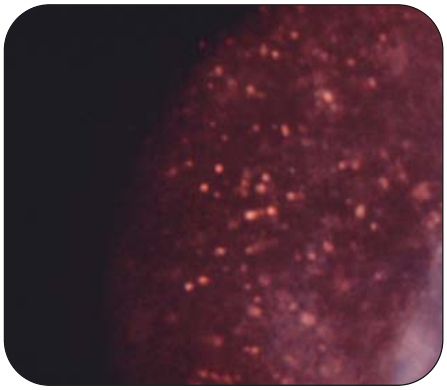

Fig. 11.10

Tumour cells

Endophytic retinoblastoma

Signs

- A friable lesion may seed tumour cells into the vitreous (**Fig. 11.11**).

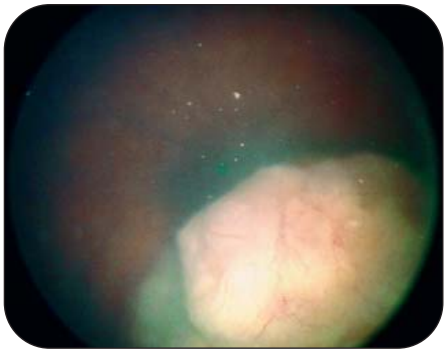

Fig. 11.11

Primary intraocular lymphoma

Signs

- Vitreous infiltration by clumps or sheets of tumour cells (**Fig. 11.12**).

Look for

- Subretinal lymphoma (**Fig. 11.13**).
- CNS disease (**Fig. 11.14**).

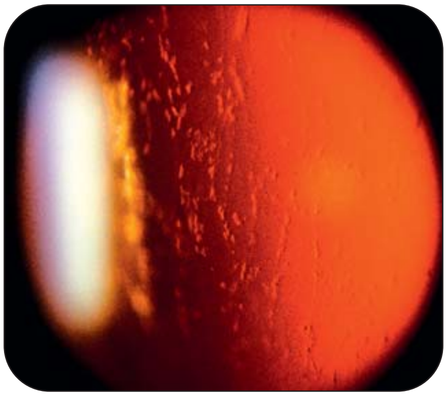

Fig. 11.12

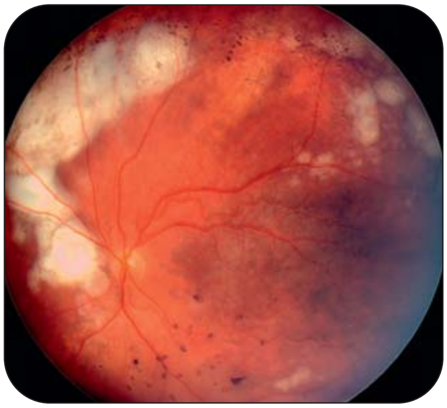

Fig. 11.13

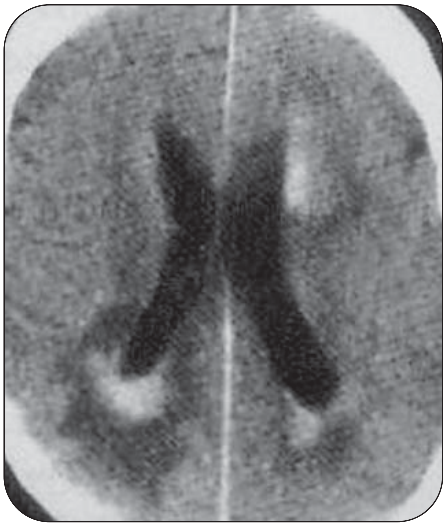

Fig. 11.14

VITREOUS OPACITIES

Common floaters

Innocuous floaters

Signs

- Muscae volitantes are bilateral, minute, multiple worm-like opacities best seen by the patient against a white or bright background.
- Larger opacities are characteristic of degeneration of the vitreous gel and are common in myopes.

Associated with acute posterior vitreous detachment

Signs

- Multiple cobwebs are condensations of collagen fibres within the collapsed vitreous gel.
- Weiss ring is a discrete ring-like opacity located on the posterior vitreous face consisting of glial tissue detached from the margin of the optic disc (**Fig. 11.15**).

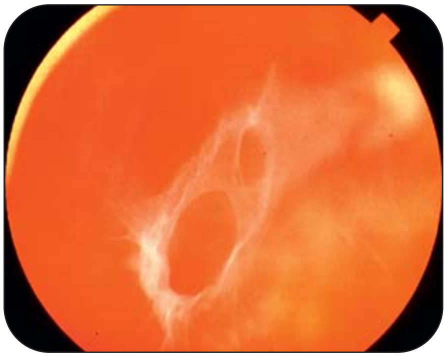

Fig. 11.15

Inflammatory

Cotton balls

Causes

- Intermediate uveitis (see Fig. 11.3).
- Sarcoidosis – usually seen inferiorly (**Fig. 11.16**).
- Candidiasis – cotton balls composed of micro-abscesses (**Fig. 11.17**).

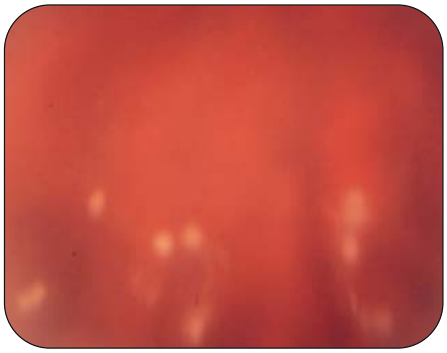

Fig. 11.16

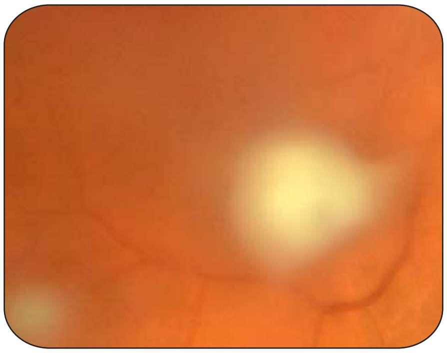

Fig. 11.17

Other

- Stringy opacities (**Fig. 11.18**) are caused by alterations in the vitreous gel itself.
- Coarse opacities (**Fig. 11.19**) are caused by severe tissue destruction.

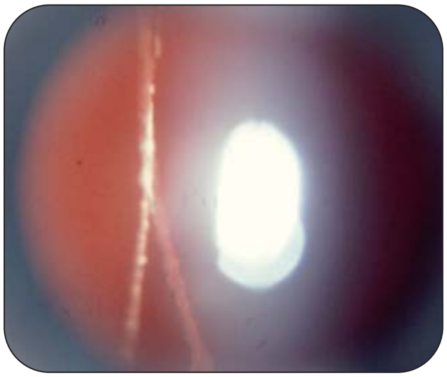

Fig. 11.18

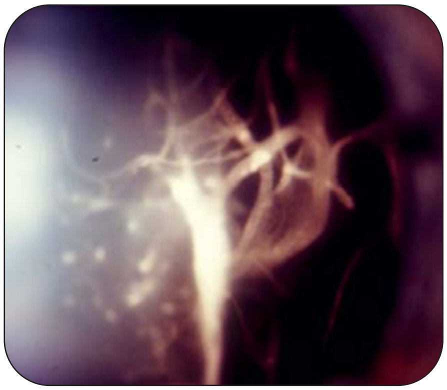

Fig. 11.19

Iridescent particles

Asteroid hyalosis (Benson disease)

Definition

- A common, usually unilateral, innocuous condition which is thought to occur with increased frequency in diabetics.

Signs

- Numerous, tiny, yellow-white spherical particles throughout the vitreous (**Fig. 11.20**).

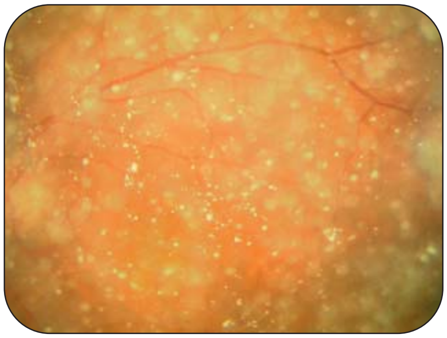

Fig. 11.20

Synchysis scintillans

Definition

- An uncommon, usually unilateral condition which occurs in eye with previous large vitreous haemorrhages.

Signs

- Numerous, golden-brown, flat refractive particles; in contrast to asteroid hyalosis, the opacities tend to sediment inferiorly when the eye is immobile.

Deposits in familial amyloidosis

Signs

- Bilateral, sheet-like ('glass-wool') opacities (**Fig. 11.21**).

Look for

- Light-near dissociation.
- Lattice dystrophy type 2 (see Fig. 5.100).
- Bilateral facial palsy.
- Peripheral and autonomic polyneuropathy.

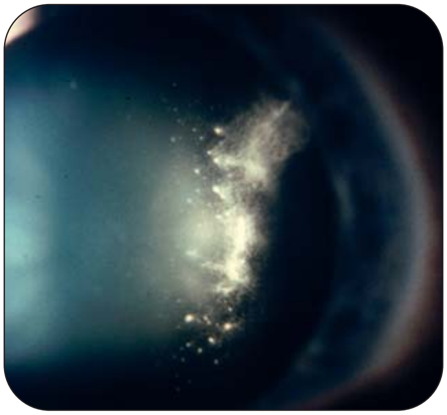
Fig. 11.21

Miscellaneous

Solitary retinal fragments (operculum)

Signs

- An operculum consists of the torn away flap of a U-shaped retinal tear (**Fig. 11.22**).

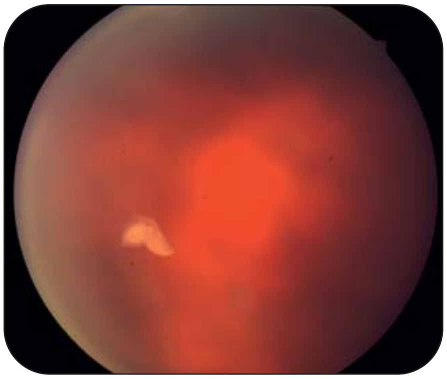
Fig. 11.22

Congenital remnants

a. *Vitreous cyst* (**Fig. 11.23**).

b. *Persistent hyaloid artery* (**Fig. 11.24**).

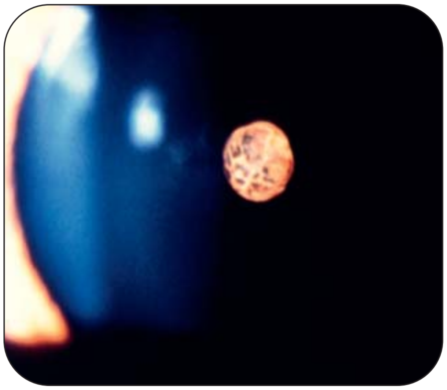
Fig. 11.23

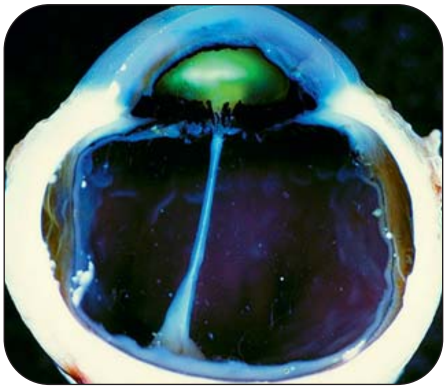
Fig. 11.24

Acquired cysts

- Cysticercosis (**Fig. 11.25**).
- Echinococcosis.

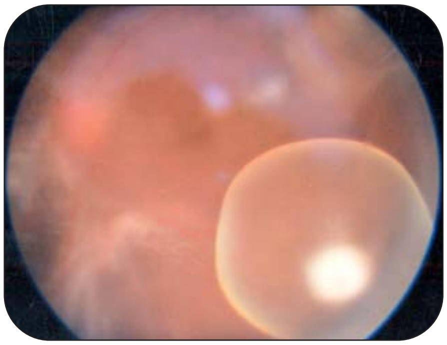

Fig. 11.25

Lens fragments

Signs

- Lens fragments and occasionally the IOL (**Fig. 11.26**) may become accidentally dislodged into the vitreous during cataract surgery.
- Rarely the lens may dislocate spontaneously in patients with hereditary ectopia lentis (see Fig. 10.11).

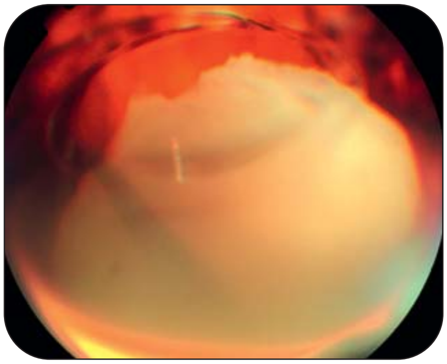

Fig. 11.26

Foreign bodies and material

a. *Metallic.*

b. *Intravitreal implants* containing ganciclovir (**Fig. 11.27**) or steroids (**Fig. 11.28**).

c. *Air and other gases* (**Fig. 11.29**).

d. *Silicone oil and heavy liquids.*

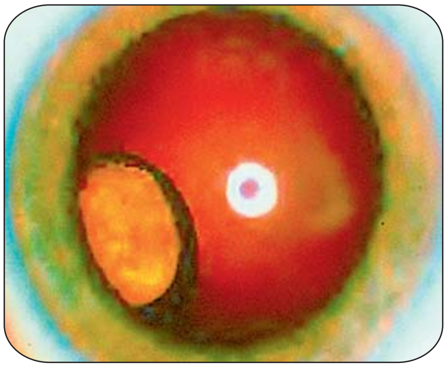

Fig. 11.27

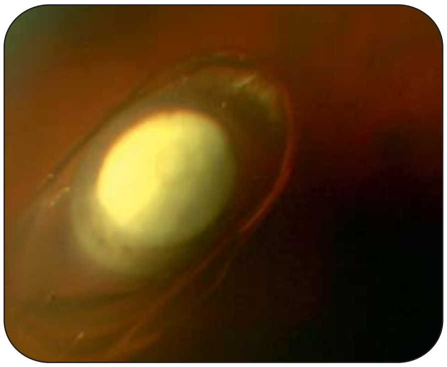

Fig. 11.28

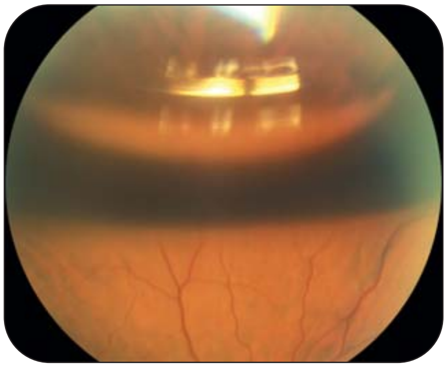

Fig. 11.29

HEREDITARY VITREORETINOPATHIES

Associated with vitreous condensations

Familial exudative vitreoretinopathy

Definition

- A rare AD condition which is similar to retinopathy of prematurity but is not associated with prematurity.

Signs

- Dragged disc vessels and macular heteropia.
- Abrupt termination of retinal vessels along a scalloped edge at the equator (**Fig. 11.30**).

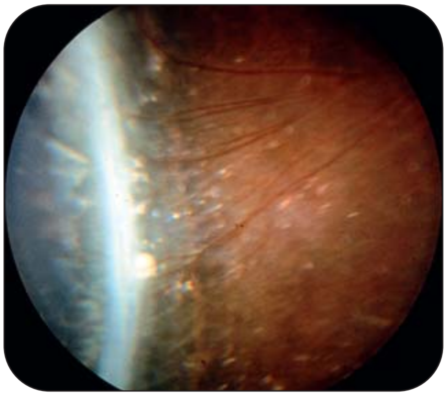

Fig. 11.30

Autosomal dominant vitreoretinochoroidopathy

Signs

- Vitritis.
- Peripheral retinal pigmentation and neovascularisation with a sharp demarcation between normal and abnormal retina.
- Cystoid macular oedema.

Autosomal dominant neovascular inflammatory vitreoretinopathy

Signs

- Vitritis.
- Peripheral retinal neovascularisation.
- Cystoid macular oedema.

Erosive vitreoretinopathy

Signs

- Nyctalopia.
- Progressive retinal pigment epithelial atrophy.

Associated with 'empty' vitreous

Vitreous liquefaction results in an optically empty cavity except for a thin layer of cortex behind the lens and avascular white membranes adherent to the retina (**Fig. 11.31**). It is seen in the following conditions.

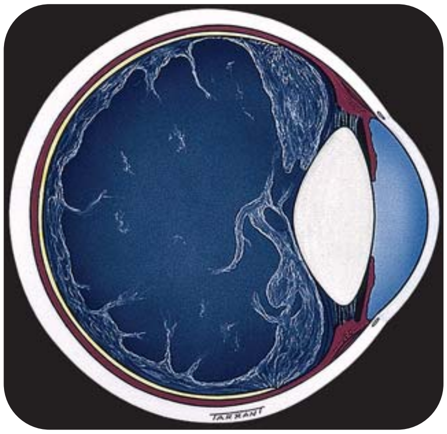

Fig. 11.31

Stickler syndrome

Definition

- An uncommon AD connective tissue disorder.

Look for

- Congenital high myopia.
- Lattice-like radial perivascular pigmentation and sclerosis (**Fig. 11.32**).
- Retinal detachment caused by giant tears (**Fig. 11.33**).
- Midfacial hypoplasia (**Fig. 11.34**).
- Arthritis, cleft palate and Robin sequence.

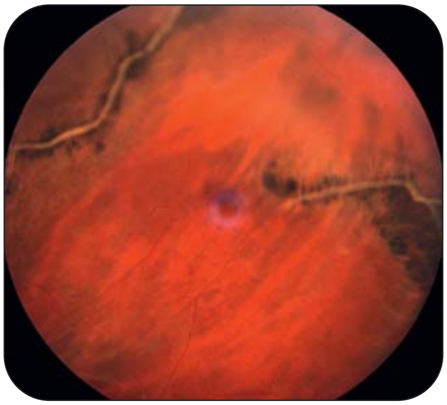

Fig. 11.32

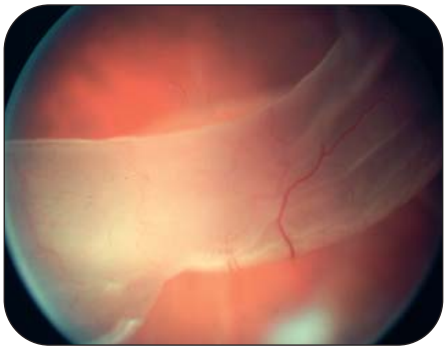

Fig. 11.33

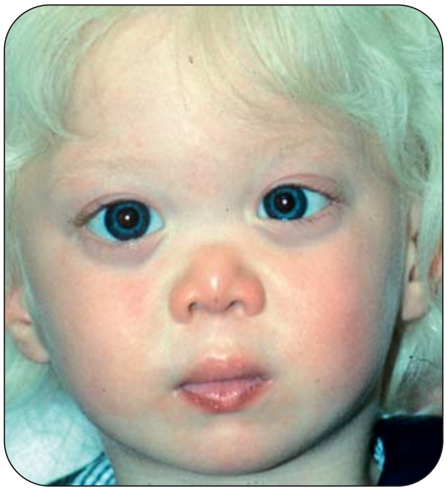

Fig. 11.34

Wagner disease

Definition

- A very rare AD condition without systemic features.

Look for

- Congenital low myopia.
- Preretinal equatorial greyish-white membranes and progressive chorioretinal atrophy (**Fig. 11.35**).
- Cataract and retinal detachment.

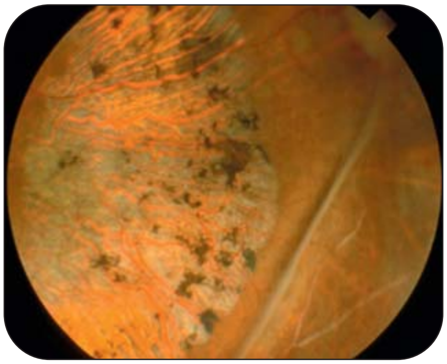

Fig. 11.35

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Optic nerve head 12

ABNORMAL DISC SIZE 251

Small 251

- *Hypermetropia* 251
- *Tilted disc* 251
- *Hypoplastic disc* 252

Large 253

- *High myopia* 253
- *Congenital pit* 253
- *Coloboma* 253
- *Morning glory anomaly* 254
- *Megalopapilla* 254
- *Combined coloboma and pit* 255

LARGE CUP 255

- *Large physiological excavation* 255
- *Glaucomatous cupping* 256
- *Megalopapilla* 256

OPTIC ATROPHY 256

- *Primary* 256
- *Secondary* 257
- *Consecutive* 257
- *Foster Kennedy syndrome* 258

DISC SWELLING AND ELEVATION 258

Unilateral 258

- *Central retinal vein occlusion* 258
- *Papillophlebitis (optic disc vasculitis)* 258
- *Non-arteritic anterior ischaemic optic neuropathy* 259
- *Arteritic anterior ischaemic optic neuropathy* 259
- *Papillitis* 259
- *Neuroretinitis* 260
- *Other causes* 260

Bilateral 260

- *Buried optic disc drusen* 260
- *Papilloedema* 261
- *Malignant (accelerated) hypertension* 263
- *Diabetic papillopathy* 263
- *Other causes* 263

VASCULAR ABNORMALITIES 264

Congenital 264

- *Pre-papillary loop* 264
- *Bergmeister papilla* 264

Acquired 264

- *Collaterals* 264
- *New vessels* 265
- *Optico-ciliary shunts* 265
- *Haemorrhage* 265

TUMOURS AND GRANULOMAS 266

- *Haemangioblastoma (capillary haemangioma)* 266
- *Cavernous haemangioma* 266
- *Melanocytoma* 266
- *Astrocytoma* 267
- *Combined hamartoma of the retina and RPE* 267
- *Choroidal melanoma* 268
- *Sarcoid granuloma* 268
- *Toxocara granuloma* 268

‘DRAGGED’ DISC 269

- *Retinopathy of prematurity* 269
- *Familial exudative vitreoretinopathy (Criswick–Schepens syndrome)* 269
- *Peripheral toxocara granuloma* 269
- *Combined hamartoma of the retina and RPE* 270
- *Persistent posterior fetal vasculature* 270

JUXTAPAPILLARY LESIONS 270

Congenital 270

- *Coloboma* 270
- *Opaque nerve fibres* 271
- *Congenital hypertrophy of the RPE* 271

Degenerative 271

- *Glaucomatous chorioretinal atrophy* 271
- *Myopic chorioretinal atrophy* 272
- *Angioid streaks* 272
- *Choroidal neovascularisation* 272

Inflammatory 273

- *Presumed ocular histoplasmosis syndrome* 273
- *Toxoplasmosis* 273
- *Toxocara granuloma* 273
- *Serpiginous choroidopathy* 274

Neoplastic 274

- *Choroidal osteoma (osseous choristoma)* 274
- *Choroidal melanoma* 274

ABNORMAL DISC SIZE

Small

Hypermetropia

Definition

- A very common, usually bilateral, condition associated with reduced axial length and smaller than normal scleral canal.

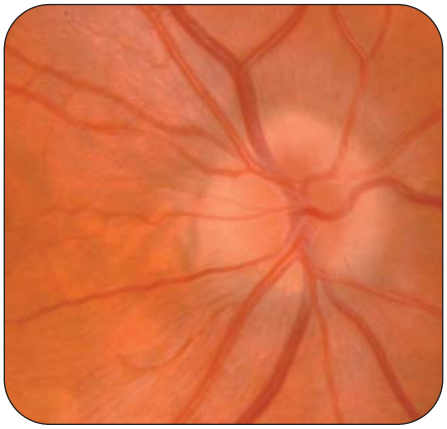

Fig. 12.1

Signs

- Slightly elevated pink disc with a small or absent cup (**Fig. 12.1**).
- Crowding of central retinal vessels.
- Absence of capillary dilatation on the disc.

Look for

- Chorioretinal folds (rare).
- Shallow anterior chamber.

Differential diagnosis

- Early papilloedema.
- Buried drusen.

Tilted disc

Definition

- A common congenital condition frequently associated with myopia.

Signs

- Oval or D-shaped disc in which the axis is most frequently directed obliquely (**Fig. 12.2**) but may be horizontal or nearly vertical (**Fig. 12.3**).

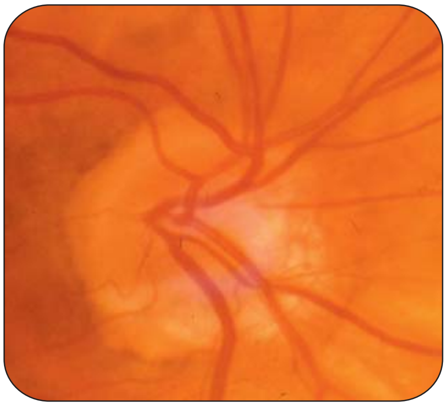

Fig. 12.2

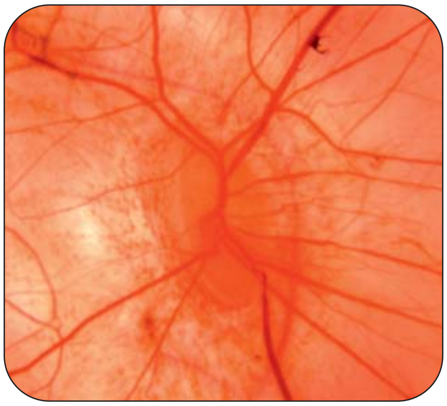

Fig. 12.3

- Situs inversus, in which the temporal vessels deviate nasally before turning temporally (**Fig. 12.4**).
- Hypopigmentation of the inferonasal fundus (**Fig. 12.5**).

Look for

- Temporal visual field defects are often present but visual acuity is usually normal.

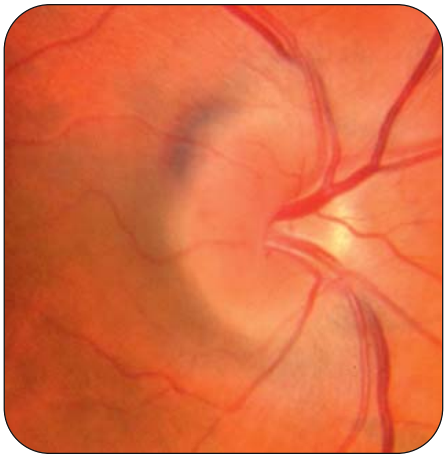

Fig. 12.4

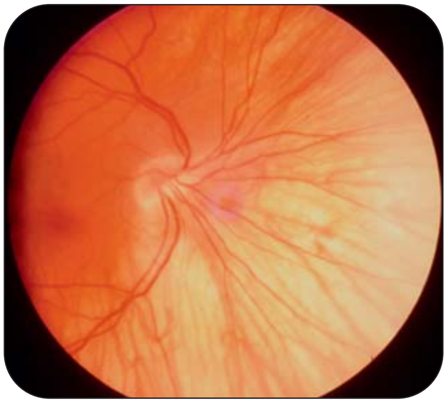

Fig. 12.5

Hypoplastic disc

Definition

- A congenital, unilateral or bilateral, condition that may be associated with a variety of ocular and systemic conditions (Table 12.1).

Signs

- Small disc surrounded by a halo of pigmentary change (double-ring sign – **Fig. 12.6**).
- Blood vessels have normal calibre although they are often tortuous (**Fig. 12.7**).
- In some cases only a part of the disc is hypoplastic (**Fig. 12.8**).

Table 12.1 **Associations of optic disc hypoplasia**

1. **Ocular**
   - Albinism and foveal hypoplasia
   - Axial myopia
   - Microphthalmos
   - Aniridia
   - Persistent hyaloid artery
2. **Maternal**
   - Drug ingestion during pregnancy (e.g. phenytoin, quinine, LSD, steroids)
   - Intrauterine infections
   - Diabetes mellitus
3. **Neurological**
   - Isolated pituitary dysfunction
   - Partial or complete absence of the septum pellucidum
   - De Morsier syndrome
   - Basal encephalocele
   - Posterior fossa cyst
   - Anencephaly
   - Hydranencephaly
   - Apert syndrome

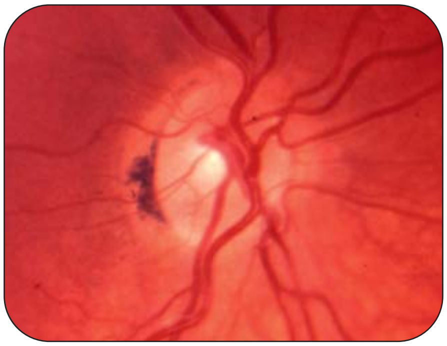

Fig. 12.6

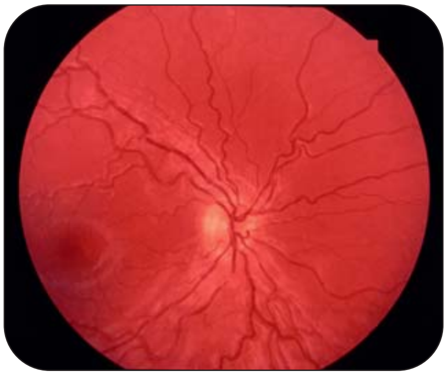

Fig. 12.7

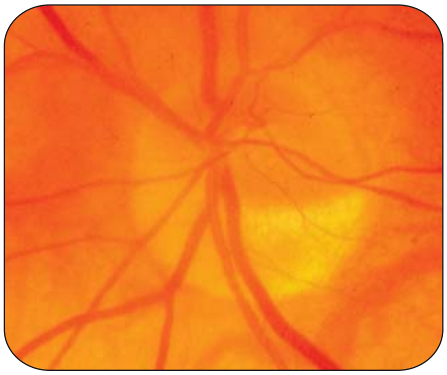

Fig. 12.8

Large

High myopia

Definition

- Refractive error more than –6D and axial length more than 26 mm with a corresponding larger than normal scleral canal.

Signs

- The appearance of the large disc may be exaggerated by surrounding chorioretinal atrophy (**Fig. 12.9**).

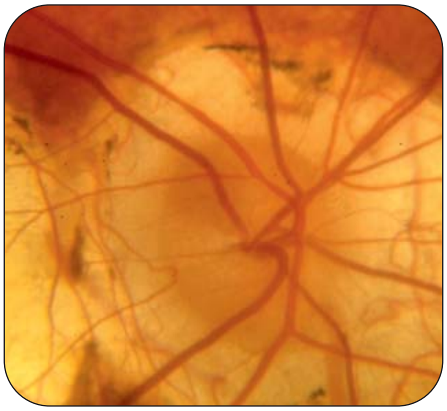

Fig. 12.9

Congenital pit

Definition

- An isolated, usually unilateral, congenital, condition that is associated with mild enlargement of the optic nerve head.

Signs

- The pit is a round or oval depression of variable size, most frequently involving the temporal margin of the disc (**Fig. 12.10**).

Look for

- Serous macular schisis and detachment (**Fig. 12.11**) that eventually develops in about 30% of cases.

Differential diagnosis of macular detachment

- Central serous retinopathy.

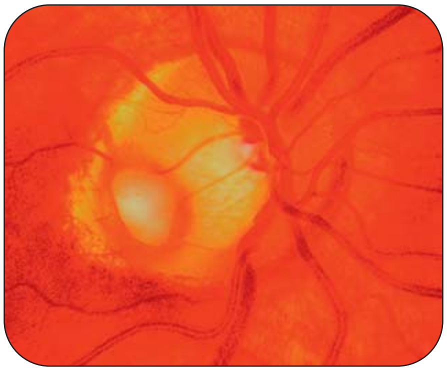

Fig. 12.10

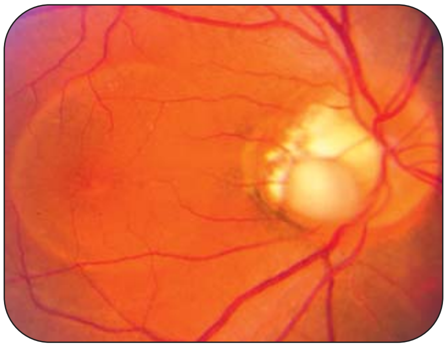

Fig. 12.11

Coloboma

Definition

- A unilateral or bilateral, congenital, condition caused by failure of fusion of the fetal fissure that may be associated with a variety of ocular and systemic anomalies (Table 12.2).

Signs

- The disc contains a partial inferior excavation that has a glistening appearance (**Fig. 12.12**).

Differential diagnosis

- Mild cases may be mistaken for glaucomatous cupping, but the splayed appearance of the central blood through the colobomatous area gives the clue.

Table 12.2 **Associations of optic disc coloboma**

1. **Ocular**
   - Other colobomas
   - Serous macular detachment
   - Microphthalmos with cyst
   - Persistent hyaloid artery
   - Retinal dysplasia
2. **Chromosomal disorders**
   - Patau syndrome (trisomy 13)
   - Edward syndrome (trisomy 18)
   - Cat-eye syndrome (trisomy 22)
3. **Brain malformations**
   - Anencephaly
   - Agenesis of corpus callosum
   - Basal encephalocele
4. **Other syndromes**
   - CHARGE
   - Meckel–Gruber
   - Goltz
   - Lenz microphthalmos
   - Aicardi
   - Hallermann–Streiff–François
   - Clefting
   - Goldenhar

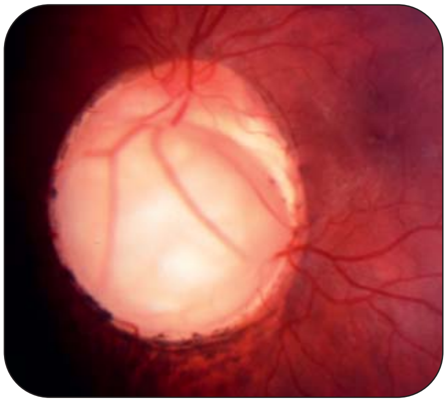

Fig. 12.12

Morning glory anomaly

Definition

- A usually unilateral, congenital, condition that may be associated with a variety of ocular and systemic anomalies (Table 12.3).

Signs

- Large disc with a core of white tissue occupying the central area (**Fig. 12.13**).
- Blood vessels emerge from the edge of the disc in a spoke-like manner.
- The lesion itself is surrounded by an elevated annulus of chorioretinal pigmentary disturbance.

Table 12.3 **Associations of morning glory anomaly**

1. **Ocular**
   - Aniridia
   - Cataract
   - Lens coloboma
   - Persistent primary fetal vasculature
   - Foveal hypoplasia
   - Serous macular detachment
2. **Uncommon systemic**
   - Hare lip and cleft palate
   - Cranial vault anomalies
   - Basal encephalocele
   - Absent corpus callosum

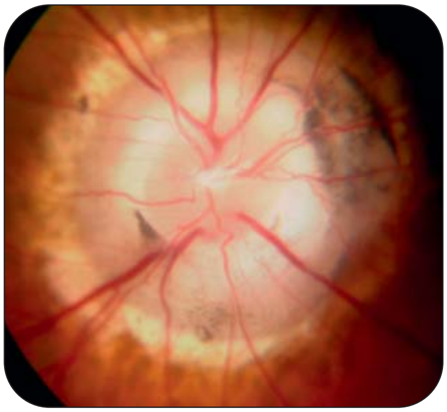

Fig. 12.13

Megalopapilla

Definition

- A very rare usually bilateral congenital condition in which the disc diameter is 2.1 mm or more.

Signs

- Large cup–disc ratio with a round or horizontally oval cup without vertical notching (**Fig. 12.14**).
- Reduced distance between the temporal edge of the disc and the foveola by approximately one disc diameter.

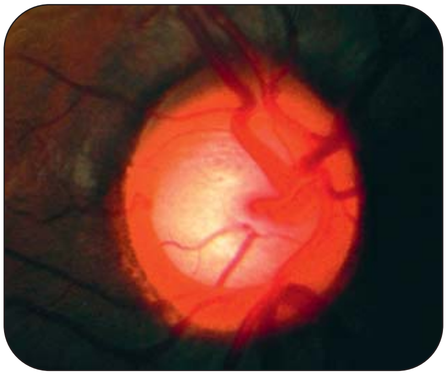

Fig. 12.14

Combined coloboma and pit

Signs

- Temporal pit and an inferior coloboma is an extremely rare anomaly (**Fig. 12.15**).

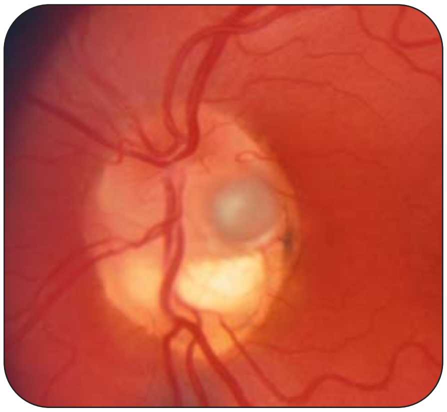

Fig. 12.15

LARGE CUP

Large physiological excavation

Signs

- Large cup with a pink neuroretinal rim with absence of notching and bayoneting of blood vessels.
- The inferior neuroretinal rim is the broadest followed by the superior, nasal and temporal (‘ISNT’ rule – **Fig. 12.16**).
- The striations of the peripapillary nerve fibre layer can be seen up to the disc margin.

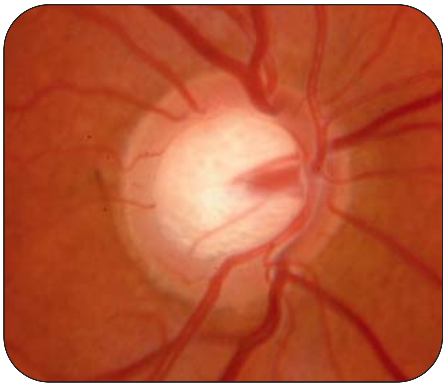

Fig. 12.16

Glaucomatous cupping

Signs

- Large cup with notching and thinning of the neuroretinal rim, and bayoneting of blood vessels (**Fig. 12.17**).
- 'ISNT' rule is broken.
- Disc pallor and peripapillary atrophy.
- Retinal nerve fibre layer defects best seen with red-free light (**Fig. 12.18**).

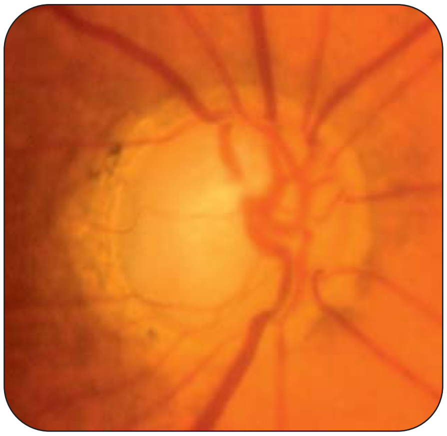

Fig. 12.17

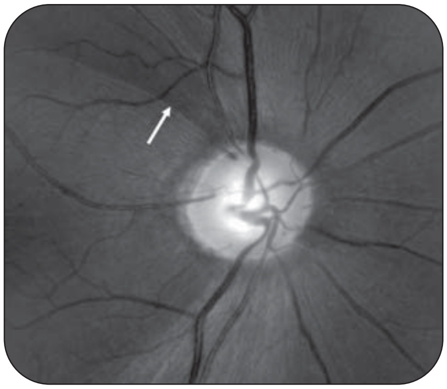

Fig. 12.18

Megalopapilla

- See above.

OPTIC ATROPHY

Primary

Pathogenesis

- A process affecting the visual pathways from the retrolaminar portion of the optic nerve to the lateral geniculate nucleus (Table 12.4).

Signs

- White, flat disc with a clearly delineated outline (**Fig. 12.19**).
- Reduction in the number of small vessels on the disc.
- Attenuation of the peripapillary vessels.
- Thinning of the retinal nerve fibre layer.

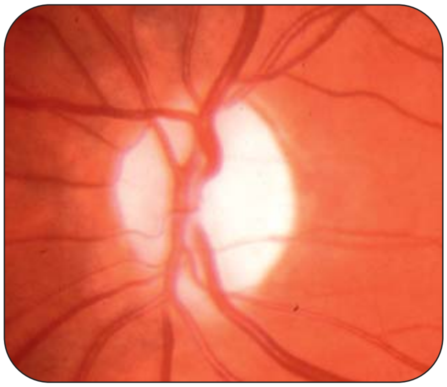

Fig. 12.19

Table 12.4 **Causes of primary optic atrophy**

1. **Optic neuritis**
    - Multiple sclerosis
    - Devic disease
    - Schilder disease
2. **Compression**
    - Bony
    - Pituitary tumours
    - Optic nerve tumours
    - Aneurysm
    - Orbital cellulitis
3. **Primary hereditary optic atrophies**
    - Kjer syndrome (AD) presents in 1st or 2nd decades
    - Behr syndrome (AR) presents in the 1st decade
    - Wolfram syndrome (AR) – DIDMOAD = diabetes insipidus, diabetes mellitus, optic atrophy, and deafness; presents 1st or 2nd decades
    - Leber hereditary optic neuropathy (mitochondrial) presents in the 2nd to 4th decades
4. **Hereditary neurological disorders – present during the 1st decade**
    - Friedreich ataxia (AR)
    - Charcot–Marie–Tooth disease (XL-D)
    - Adrenoleukodystrophies (may be XL-R or AR)
    - Cerebellar ataxia type I (AD)
5. **Toxic**
    - Methanol
    - Ethambutol
    - Isoniazid
6. **Nutritional deficiency**
    - Thiamine (tobacco-alcohol amblyopia and beriberi)
    - Vitamin B12 (pernicious anaemia)
    - Niacin (pellagra)
7. **Trauma**
    - With or without optic nerve transection
    - Retrobulbar haemorrhage

Secondary

Pathogenesis

- Long-standing swelling of the optic nerve head.

Signs

- White or dirty grey slightly raised disc with poorly delineated margins (**Fig. 12.20**).
- Reduction in number of small vessels on the disc.
- Surrounding 'water marks'.

Causes

- Chronic papilloedema.
- Anterior ischaemic optic neuropathy.
- Papillitis.

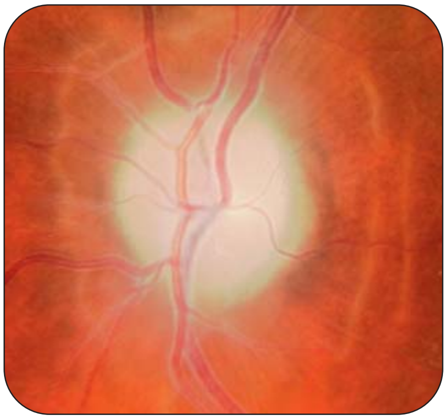

Fig. 12.20

Consecutive

Pathogenesis

- Diseases of the inner retina or its blood supply (Table 12.5).

Signs

- The cause is usually obvious from examination of the fundus such as retinitis pigmentosa (**Fig. 12.21**) and old vasculitis associated with Behçet disease (**Fig. 12.22**).

Table 12.5 **Fundus changes associated with consecutive optic atrophy**

1. **Vascular attenuation or sheathing**
   - Old occlusion of the central retinal artery or ophthalmic artery
   - Retinitis pigmentosa
   - Toxic optic neuropathy
   - Old vasculitis as in Behçet disease
2. **Macular lesions giving rise to temporal disc pallor**
   - Toxoplasmosis
   - Cone dystrophy
3. **Chorioretinal atrophy**
   - Degenerative myopia
   - Extensive panretinal photocoagulation
4. **Diffuse retinal necrosis**
   - Cytomegalovirus retinitis
   - Acute retinal necrosis
   - Progressive outer retinal necrosis

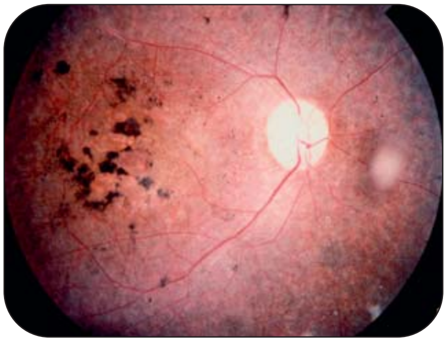

Fig. 12.21

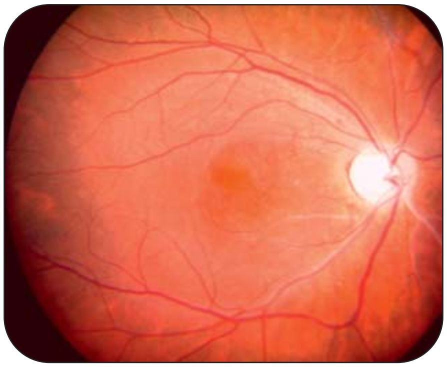

Fig. 12.22

Foster Kennedy syndrome

a. *True F-K syndrome* is a very rare condition that is classically caused by a frontal-space-occupying lesion that causes ipsilateral compressive optic atrophy and contralateral papilloedema.

b. *Pseudo-F-K syndrome* is more common and is caused by anterior ischaemic optic neuropathy in which active disease in one eye (**Fig. 12.23A**) is associated with contralateral atrophy in the other (**Fig. 12.23B**) due to a previous episode.

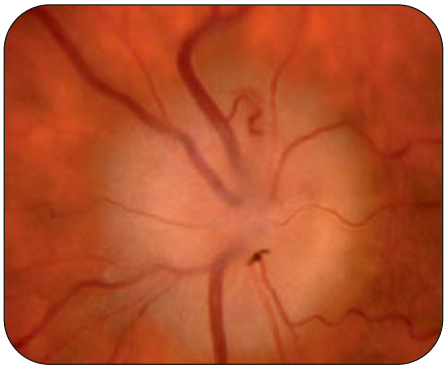

Fig. 12.23A

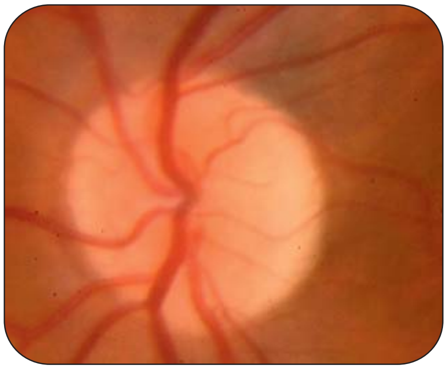

Fig. 12.23B

DISC SWELLING AND ELEVATION

Unilateral

Central retinal vein occlusion

Definition

- A common, usually unilateral, condition that may be associated with severe visual impairment.

Signs

- Mild hyperaemic disc swelling.
- Venous engorgement, widespread retinal haemorrhages and variable cotton-wool spots (**Fig. 12.24**).

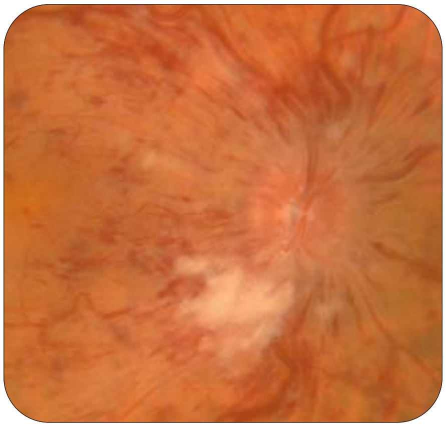

Fig. 12.24

Papillophlebitis (optic disc vasculitis)

Definition

- An uncommon condition that may be a variant of central retinal vein occlusion although it affects younger healthy individuals and has a better prognosis.

Signs

- Severe hyperaemic disc swelling.
- Peripapillary haemorrhages, and variable venous engorgement and cotton-wool spots (**Fig. 12.25**).

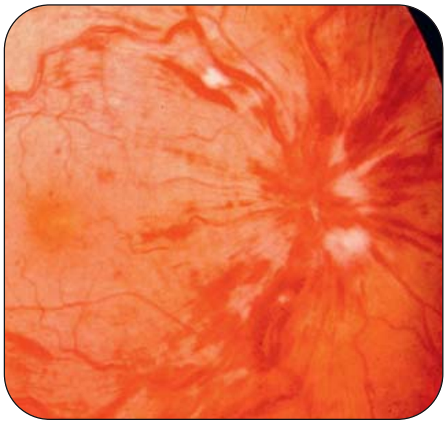

Fig. 12.25

Non-arteritic anterior ischaemic optic neuropathy

Definition

- Infarction of the optic nerve head not associated with giant cell arteritis but with a variety of predisposing factors (Table 12.6).

Signs

- Diffuse or sectoral hyperaemic disc swelling often associated with small peripapillary haemorrhages (**Fig. 12.26**).

Table 12.6 **Predispositions to non-arteritic anterior ischaemic optic neuropathy**

1. **Ocular**
   - Small optic cup
   - Optic disc drusen
2. **Systemic**
   - Hypertension
   - Hyperlipidaemia
   - Migraine
   - Acute blood loss

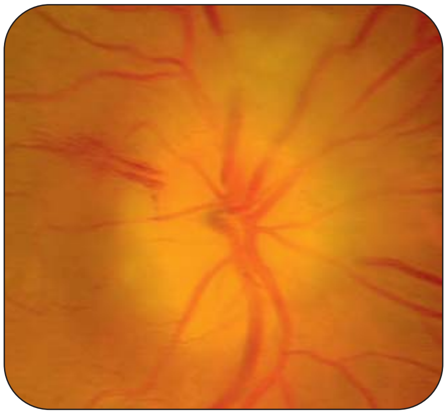

Fig. 12.26

Arteritic anterior ischaemic optic neuropathy

Definition

- Infarction of the optic nerve head caused by giant-cell arteritis.

Signs

- Pale diffuse disc swelling that may be associated with small peripapillary haemorrhages (**Fig. 12.27**).

Look for

- Polymyalgia rheumatica.
- Headache.
- Jaw claudication.
- Fever.
- Tender, nodular temporal arteries.
- Scalp necrosis, in very severe cases.

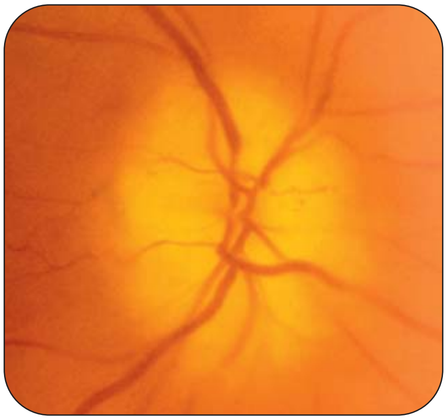

Fig. 12.27

Papillitis

Definition

- Intraocular form of optic neuritis that may be associated with a variety of infectious and non-infectious conditions (Table 12.7).
- Children may have bilateral involvement.

Signs

- Hyperaemic disc swelling is often florid and associated with peripapillary haemorrhages (**Fig. 12.28**).

Table 12.7 **Causes of papillitis**

1. **Idiopathic**
2. **Parainfectious**
   - Viral infections (e.g. measles, mumps, chickenpox and rubella)
   - Glandular fever
   - Following immunisation
3. **Infectious**
   - Sinusitis-related
   - Cat-scratch fever
   - Syphilis
   - Herpes zoster
   - Lyme disease
   - Cryptococcal meningitis
4. **Non-infectious**
   - Sarcoidosis
   - Autoimmune
   - Demyelination is rare

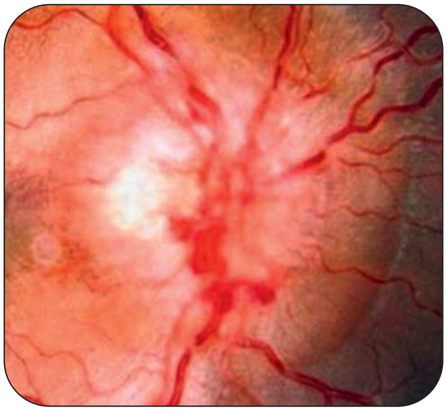

Fig. 12.28

Neuroretinitis

Definition

- Inflammation of the optic nerve and retina that may occur at any age.

Signs

- Papillitis associated with a macular star (**Fig. 12.29**).
- The macular star usually becomes more prominent as the papillitis subsides.

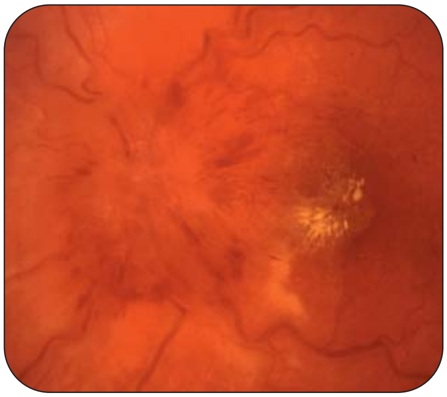

Fig. 12.29

Causes

- Idiopathic (Leber stellate neuroretinitis).
- Cat scratch fever is the most common.
- Syphilis, Lyme disease, mumps and leptospirosis.

Other causes

a. *Posterior scleritis* is a unilateral or bilateral condition that may be associated with systemic disease.

b. *Post-cataract papillopathy* is a rare ischaemic condition that occurs between 4 weeks and 15 months after uncomplicated cataract surgery.

c. *Compressive optic nerve lesions* (Table 12.8) may initially cause disc oedema and later optic atrophy.

Table 12.8 **Causes of optic nerve compression**

1. **Intraorbital**
   - Optic nerve glioma
   - Meningioma
   - Thyroid ophthalmopathy
   - Orbital tumours
   - Orbital cellulitis
2. **Intracanalicular**
   - Paget disease
   - Fibrous dysplasia
   - Osteopetrosis
3. **Intracranial**
   - Meningioma
   - Pituitary adenoma

Bilateral

Buried optic disc drusen

Definition

- A common, often familial, condition caused by deposition of hyaline material within the optic nerve head that is occasionally associated with ocular and systemic disorders (Table 12.9).
- It is the most common cause of pseudo-papilloedema in children.

Table 12.9 **Associations of disc drusen**

1. **Ocular**
   - Disc haemorrhages
   - Retinal nerve fibre bundle visual field defects
   - Peripapillary choroidal neovascularisation
   - Increased incidence of anterior ischaemic optic neuropathy
   - Angioid streaks
   - Retinitis pigmentosa
2. **Systemic**
   - Alagille syndrome

Signs

- Elevated, 'lumpy'-looking disc that is not hyperaemic (**Fig. 12.30**).
- No physiological cup.
- Disc vessels are not obscured as they pass over the disc.
- Emerging vessels are more numerous than normal and show anomalous branching patterns.
- Peripapillary retinal nerve fibre layer is usually normal.
- With age, the drusen become easier to detect because they enlarge and become exposed above the disc surface (**Fig. 12.31**).
- The lesions show autofluorescence (**Fig. 12.32**) and can be detected by ultrasonography (**Fig. 12.33**) and CT (**Fig. 12.34**).

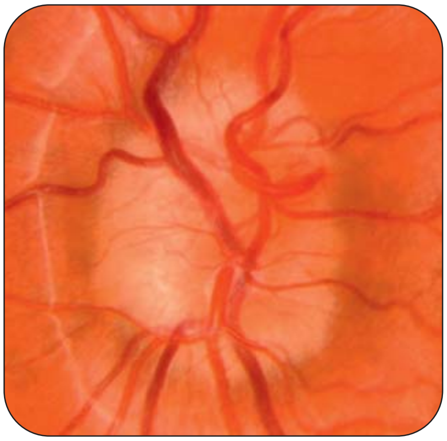
Fig. 12.30

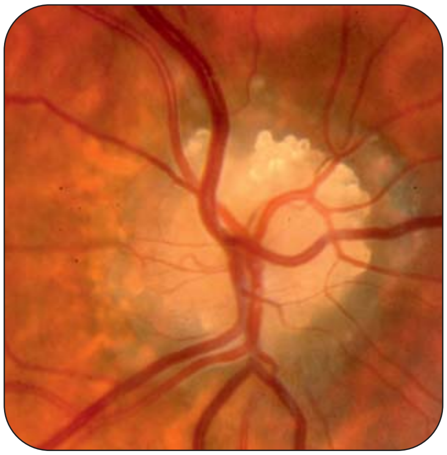
Fig. 12.31

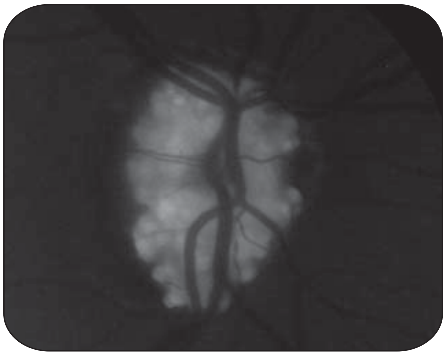
Fig. 12.32

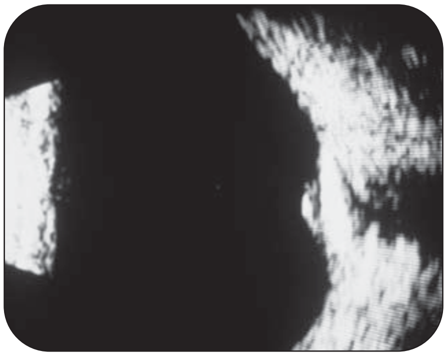
Fig. 12.33

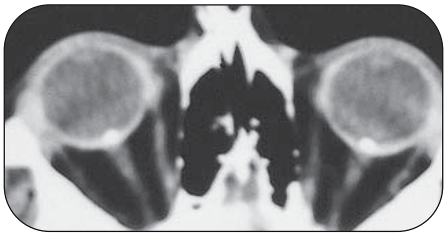
Fig. 12.34

Papilloedema

Definition

- Disc oedema secondary to raised intracranial pressure (Table 12.10) which is invariably bilateral but may be asymmetric. The signs depend on the severity and duration.

Table 12.10 **Causes of raised intracranial pressure**

1. **Space-occupying intracranial lesions**
   - Tumour
   - Haemorrhage
2. **Idiopathic intracranial hypertension (pseudotumor cerebri)**
3. **Infections**
   - Meningitis
   - Encephalitis
4. **Toxic**
   - Lead poisoning
   - Chronic hypervitaminosis A
5. **Metabolic**
   - Chronic renal failure
   - Hypercapnia
6. **Developmental**
   - Aqueduct stenosis
   - Craniodysostoses
   - Arteriovenous malformations
7. **Iatrogenic**
   - Radical neck surgery
   - Tetracycline

Signs

a. Early (**Fig. 12.35**)

- Mild disc hyperaemia and elevation with preservation of the physiological cup.
- Indistinct disc margins and peripapillary retinal nerve fibre striations.
- The nasal sector is usually involved first, followed by the superior and inferior sectors.
- Spontaneous venous pulsation is absent – this may not be significant because it is also absent in about 20% of the general population.
- Differential diagnosis – hypermetropia and buried drusen.

b. **Established** *(acute –* **Fig. 12.36***)*

- Moderate disc elevation, blurring of the entire disc margin, absence of the physiological cup and severe hyperaemia.
- Venous engorgement, flame-shaped haemorrhages and cotton-wool spots.
- Circumferential peripapillary retinal folds (Paton lines) due to fluid accumulation.
- Differential diagnosis – malignant hypertension.

c. Chronic (**Fig. 12.37**)

- Marked disc elevation of the disc without haemorrhages or cotton-wool spots.
- Telangiectasia, optico-ciliary shunts and drusen-like crystalline deposits may be present.
- A macular star is common.

d. **Atrophic** *(secondary optic atrophy –* **Fig. 12.38***)*

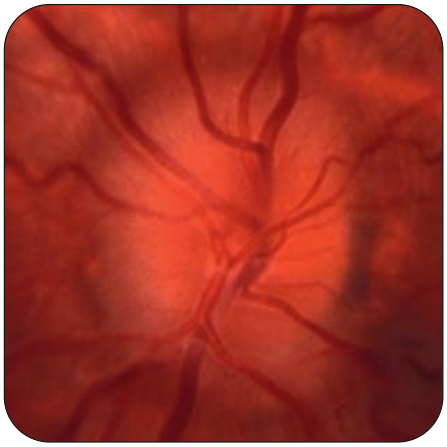
Fig. 12.35

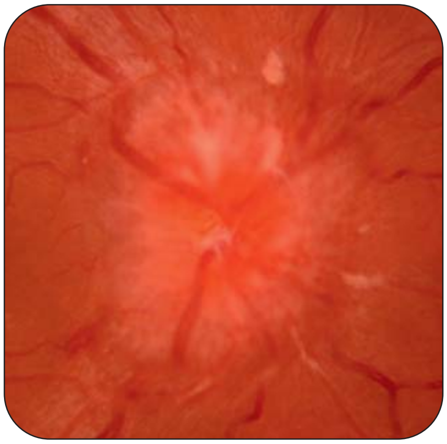
Fig. 12.36

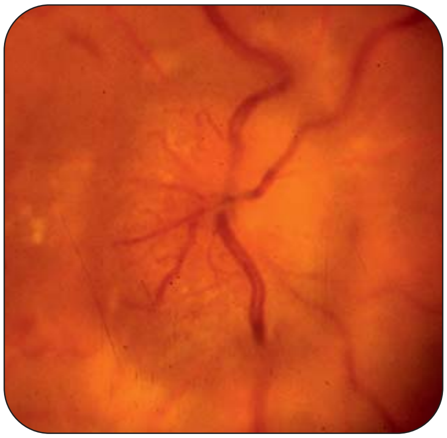
Fig. 12.37

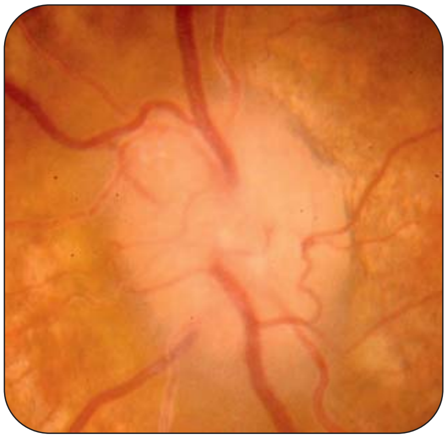
Fig. 12.38

Malignant (accelerated) hypertension

Definition

- Very high blood pressure associated with rapidly progressive end-organ damage.

Signs

- Disc swelling, flame-shaped haemorrhages, cotton-wool spots and a macular star (**Fig. 12.39**).
- According to the age of the patient there are also variable retinal hypertensive vascular changes.

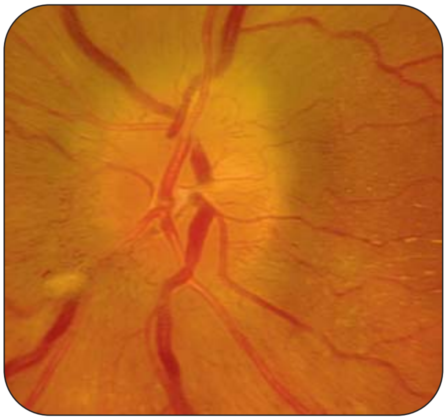

Fig. 12.39

Diabetic papillopathy

Definition

- An uncommon condition of unclear aetiology that typically affects young diabetic patients.

Signs

- Mild hyperaemic disc swelling associated with surface telangiectasia (**Fig. 12.40**).

Differential diagnosis

- Disc neovascularisation.

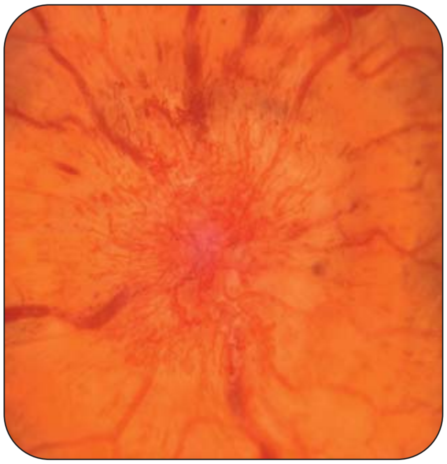

Fig. 12.40

Other causes

a. *Compressive thyroid optic neuropathy* may give rise to bilateral disc swelling in the absence of significant proptosis.

b. *Acute methanol toxicity* is a very rare condition characterised by acute bilateral visual loss and dilated pupils.

c. *Bilateral, simultaneous, arteritic anterior ischaemic optic neuropathy*.

d. *Bilateral papillitis* which typically affects children.

e. *Compromised venous drainage* may result in bilateral disc swelling in patients with cavernous sinus thrombosis or direct carotid–cavernous fistula.

VASCULAR ABNORMALITIES

Congenital

Pre-papillary loop

Signs

- A usually unilateral anomaly in which an arterial loop extends from the disc into the vitreous cavity and then back (**Fig. 12.41**).

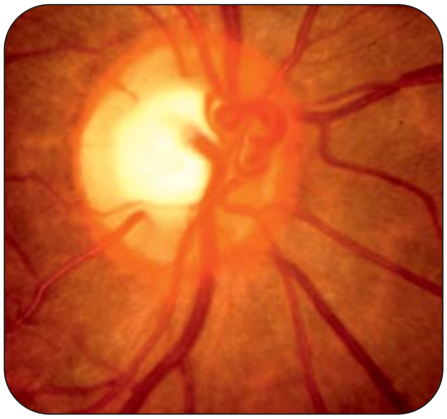
Fig. 12.41

Bergmeister papilla

Signs

- A unilateral condition characterised by a raised glial tissue on the disc (**Fig. 12.42**).

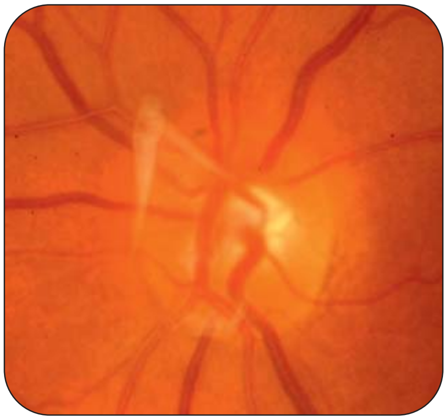
Fig. 12.42

Acquired

Collaterals

Definition

- Veno-venular shunts that develop within the framework of the existing vascular network to bypass an area of closure most frequently associated with retinal vein occlusion.

Signs

- Distended flat vessels that start and end on the disc (**Fig. 12.43**).

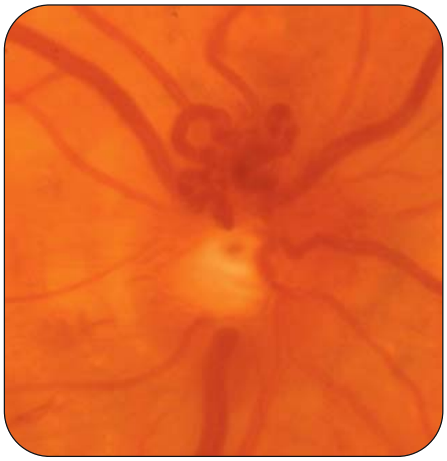
Fig. 12.43

New vessels

Definition

- New vessel formation may be unilateral or bilateral depending on the cause (see Chapter 13).

Signs

- Fine, lace-like vessels that may extend onto adjacent retina (**Fig. 12.44**).
- The vessels may be flat or elevated above the disc and may be associated with gliosis.

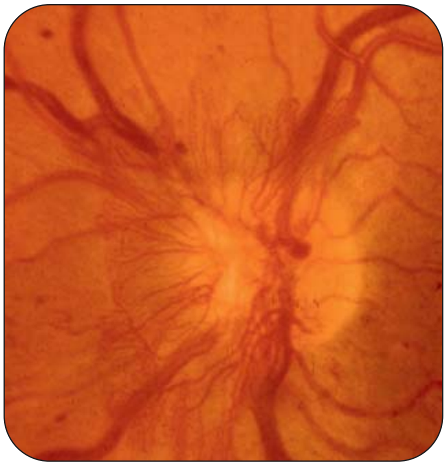

Fig. 12.44

Optico-ciliary shunts

Definition

- Usually unilateral anastomoses between the retinal and choroidal circulations.

Signs

- Slightly tortuous vessels that disappear at the retina–disc junction (**Fig. 12.45**).

Causes

- Optic nerve sheath meningioma (in 20–30% of cases).
- Optic nerve glioma (uncommon).
- Chronic papilloedema.

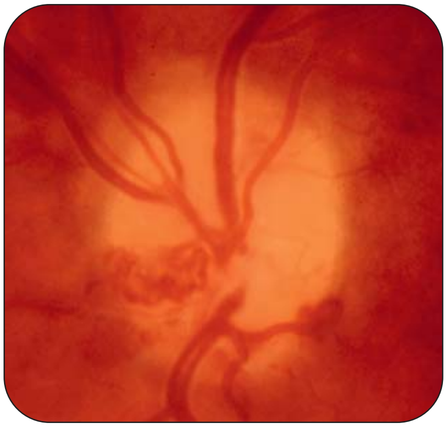

Fig. 12.45

Haemorrhage

Signs

- Peripheral flame-shaped haemorrhage that may be solitary or multiple (**Fig. 12.46**).

Table 12.11 **Causes of disc haemorrhage**

- Acute papilloedema
- Anterior ischaemic optic neuropathy
- Papillitis
- Chronic glaucoma, particularly low tension
- Infiltrative optic neuropathy
- Following optic nerve sheath decompression
- In association with acute posterior vitreous detachment
- Optic disc drusen

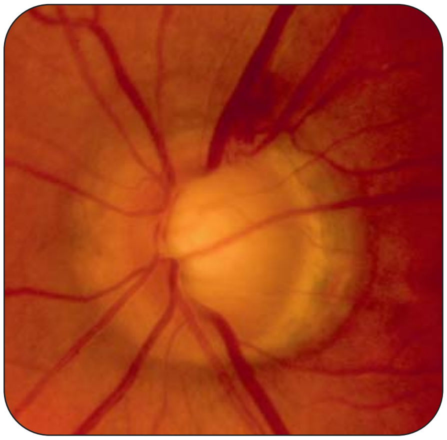

Fig. 12.46

TUMOURS AND GRANULOMAS

Haemangioblastoma (capillary haemangioma)

Definition

- A unilateral or bilateral, solitary or multiple, benign but vision-threatening tumour that more frequently involves the peripheral retina than the optic nerve head.

Signs

- Orange-red mass that may be associated with macular hard exudates (**Fig. 12.47**).

Look for

- Cerebral haemangioblastomas (**Fig. 12.48**) in von Hippel–Lindau syndrome which is present in 50% of patients with solitary lesions and in virtually all cases with multifocal tumours.

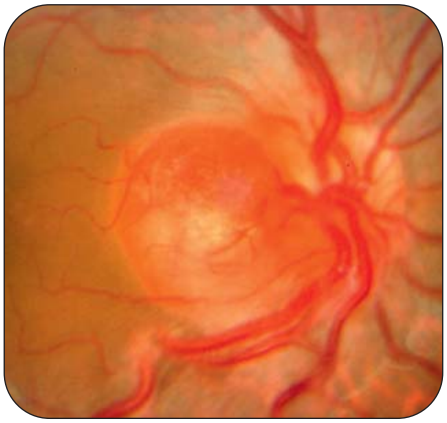

Fig. 12.47

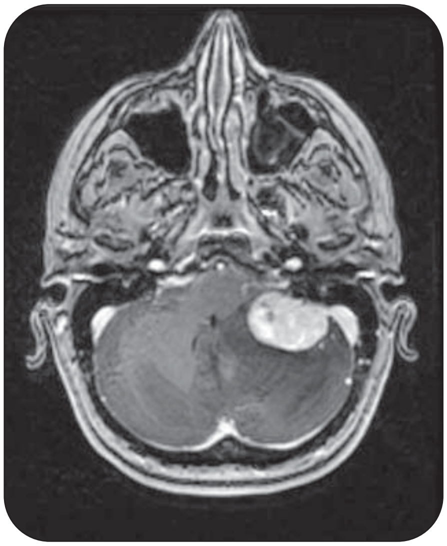

Fig. 12.48

Cavernous haemangioma

Definition

- A unilateral benign and innocuous tumour that more frequently involves the retina than the optic nerve head.

Signs

- Clumps of thin-walled saccular aneurysms filled with dark blood vessels reminiscent of a cluster of grapes (**Fig. 12.49**).

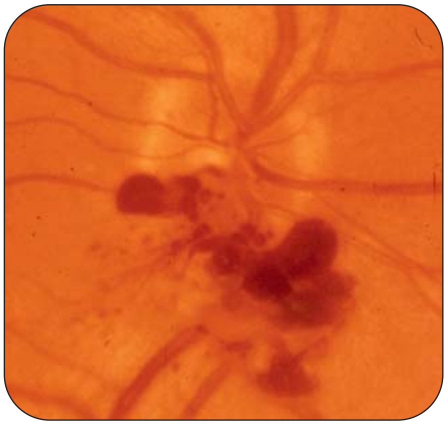

Fig. 12.49

Melanocytoma

Definition

- A unilateral, benign tumour that typically affects the optic nerve head of heavily pigmented individuals.

Signs

- A heavily pigmented lesion that may spill over into the adjacent retina (**Fig. 12.50**).

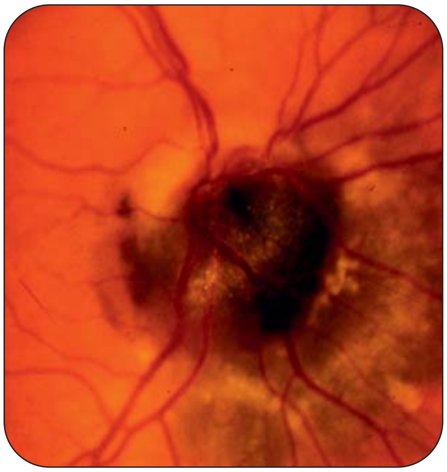

Fig. 12.50

Astrocytoma

Definition

- An innocuous, unilateral or bilateral, solitary or multiple, tumour that more frequently involves the retina than the optic nerve head. It has important systemic implications.

Signs

- Semi-translucent lesion with a mulberry-like configuration (**Fig. 12.51**).

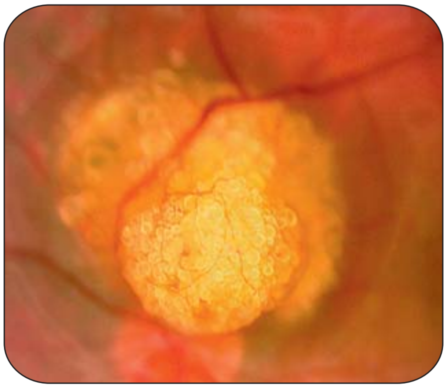

Fig. 12.51

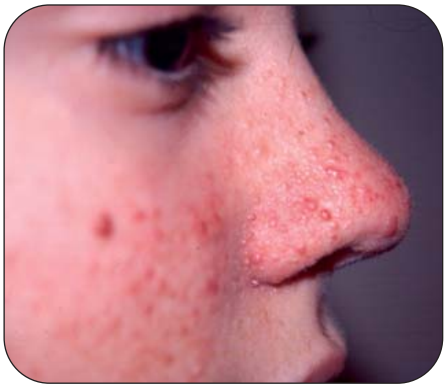

Fig. 12.52

Look for

- Adenoma sebaceum (**Fig. 12.52**) as part of tuberous sclerosis which is present in the majority of patients.

Combined hamartoma of the retina and RPE

Definition

- A benign but vision-threatening, usually unilateral malformation that typically affects males.

Signs

- A thickened epiretinal membrane overlying a dark-grey pigmented lesion associated with stress lines in the surrounding retina (**Fig. 12.53**).

Look for

- NF2 is an occasional association.

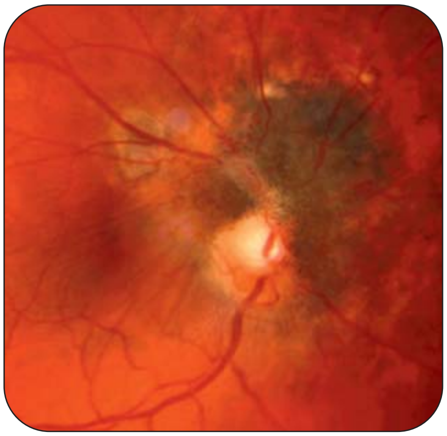

Fig. 12.53

Choroidal melanoma

Signs

- Melanoma may rarely invade the optic nerve head and cause severe visual loss (**Fig. 12.54**).

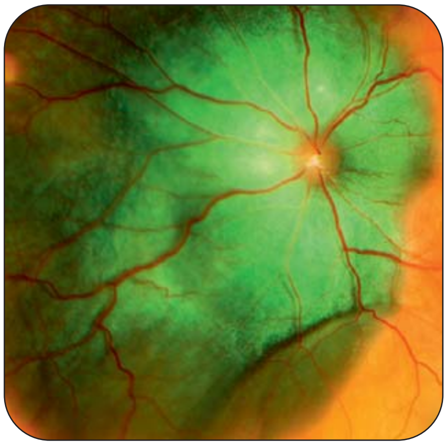

Fig. 12.54

Sarcoid granuloma

Signs

- Disc involvement (**Fig. 12.55**) is rare and does not usually affect vision.

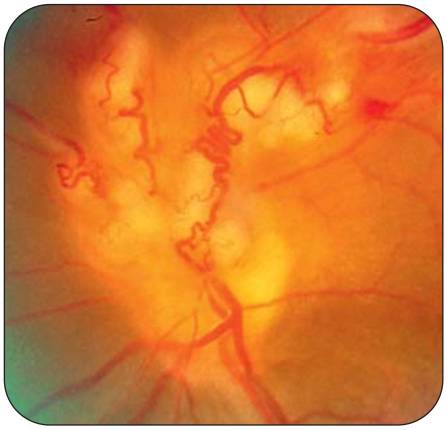

Fig. 12.55

Toxocara granuloma

Signs

- The granuloma (**Fig. 12.56**) is always unilateral and typically presents between the ages of 6 and 14 years.

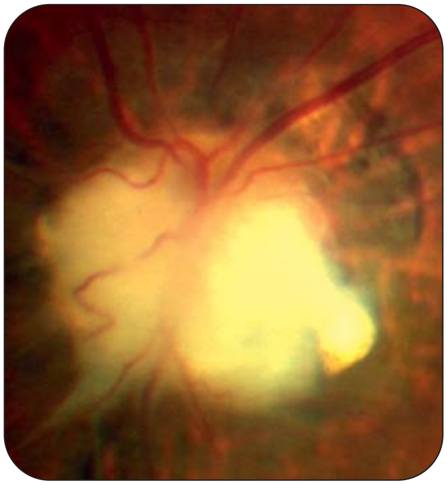

Fig. 12.56

'DRAGGED' DISC

Retinopathy of prematurity

Definition

- A bilateral proliferative retinopathy that affects premature infants of low birth weight who have been exposed to high ambient oxygen concentrations.

Signs

- Temporal disc and macula dragging occurs in advanced cicatricial disease (**Fig. 12.57**).

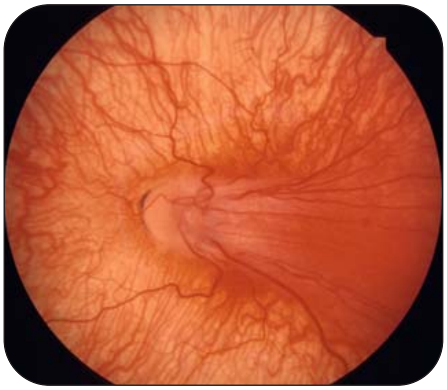

Fig. 12.57

Familial exudative vitreoretinopathy (Criswick–Schepens syndrome)

Definition

- A rare bilateral AD condition characterised by failure of vascularisation of the peripheral retina, similar to retinopathy of prematurity, but not associated with low birth weight and prematurity.

Signs

- Temporal vascular straightening and dragging of the macula and disc occur in advanced cases (**Fig. 12.58**).

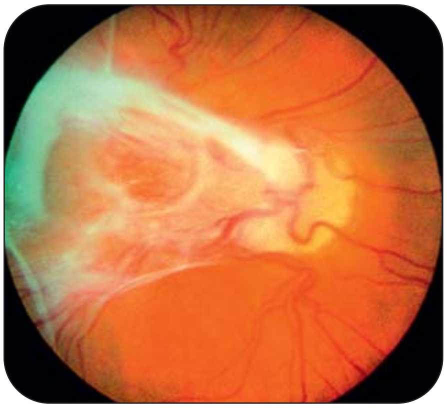

Fig. 12.58

Peripheral toxocara granuloma

Signs

- Vitreous bands extending from the lesion to the posterior fundus may cause disc and macular dragging that may involve any quadrant (**Fig. 12.59**).

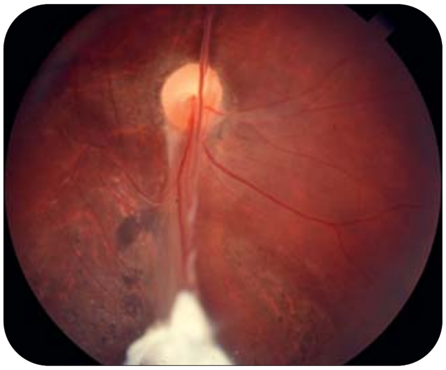

Fig. 12.59

Combined hamartoma of the retina and RPE

Signs

- Contraction may cause dragging of the optic nerve head and macula (**Fig. 12.60**).

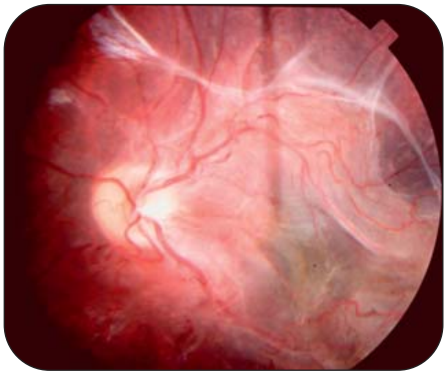

Fig. 12.60

Persistent posterior fetal vasculature

Definition

- A rare, unilateral, congenital condition that results from failure of regression of the primary vitreous.

Signs

- A retinal fold extending from ora serrata to the disc may cause dragging (**Fig. 12.61**).

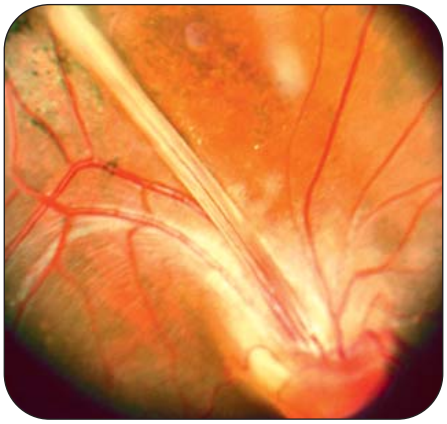

Fig. 12.61

JUXTAPAPILLARY LESIONS

Congenital

Coloboma

Signs

- A unilateral anomaly with the coloboma located inferiorly (**Fig. 12.62**).

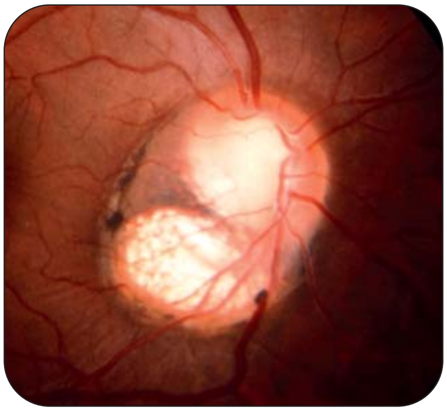

Fig. 12.62

Opaque nerve fibres

Signs

- White lesions with feathery edges which may radiate outward from the disc (**Fig. 12.63**).

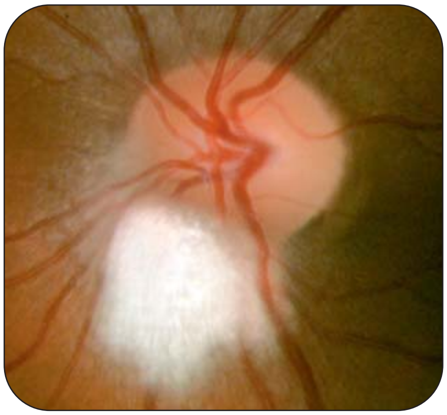

Fig. 12.63

Congenital hypertrophy of the RPE

Signs

- A juxtapapillary location is uncommon (**Fig. 12.64**).

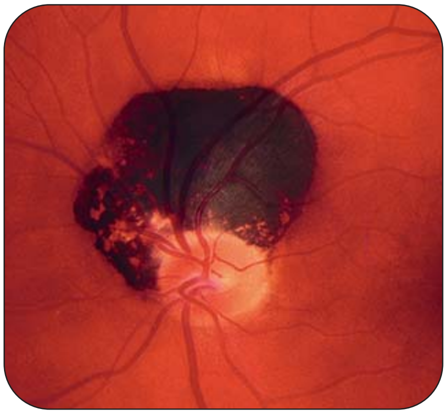

Fig. 12.64

Degenerative

Glaucomatous chorioretinal atrophy

Significance

- Occurs more frequently in primary open-angle glaucoma and tends to be more advanced in the more severely affected eye.
- It is also seen more frequent in ocular hypertension that subsequently converts to glaucoma.

Signs

- Crescent-shaped chorioretinal atrophy involving the temporal disc margin (**Fig. 12.65**).

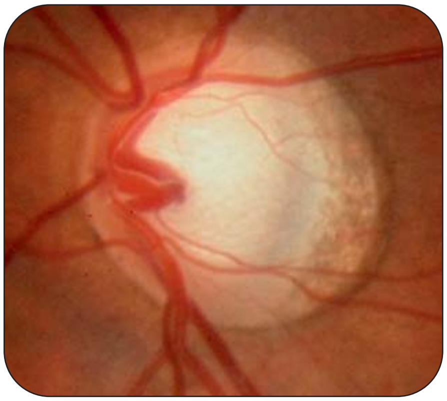

Fig. 12.65

Myopic chorioretinal atrophy

Signs

- Peripapillary chorioretinal atrophy which may also involve the posterior pole (**Fig. 12.66**).

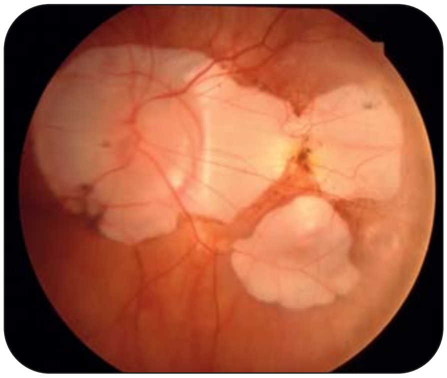

Fig. 12.66

Angioid streaks

Definition

- Crack-like dehiscences in abnormal collagenous and elastic portions of Bruch membrane.

Signs

- Dark linear lesions with serrated edges encircling the disc and then radiating outwards (**Fig. 12.67**).

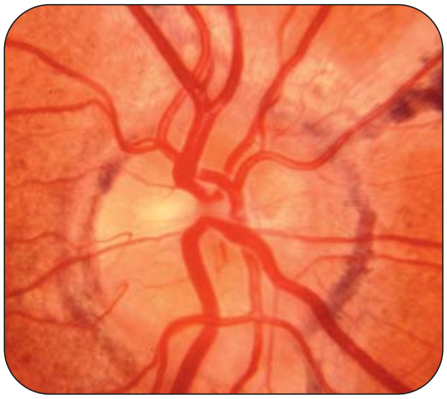

Fig. 12.67

Choroidal neovascularisation

Signs

- The neovascular membrane typically involves the temporal disc border and gives rise to oedema and haemorrhage (**Fig. 12.68**).

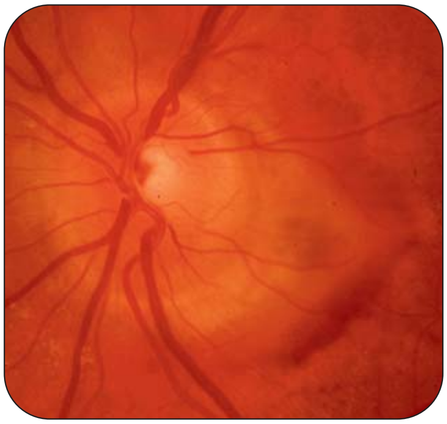

Fig. 12.68

Inflammatory

Presumed ocular histoplasmosis syndrome

Signs

- Juxtapapillary atrophy associated with multiple atrophic ‘histo’ spots in the mid-periphery and posterior pole (**Fig. 12.69**).

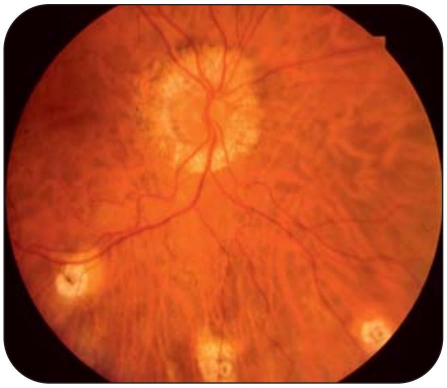

Fig. 12.69

Toxoplasmosis

Signs

- Involvement of the edge of the disc (Jensen choroiditis) is uncommon but may give rise to visual loss (**Fig. 12.70**).

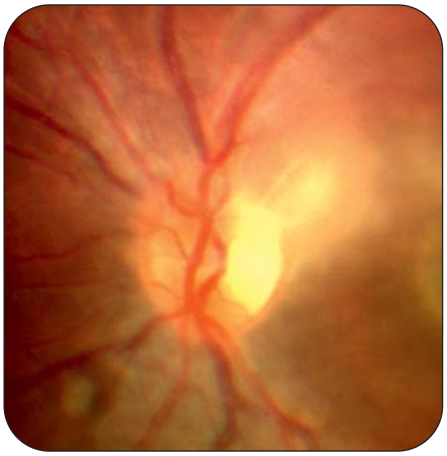

Fig. 12.70

Toxocara granuloma

Signs

- Juxtapapillary location is common (**Fig. 12.71**).

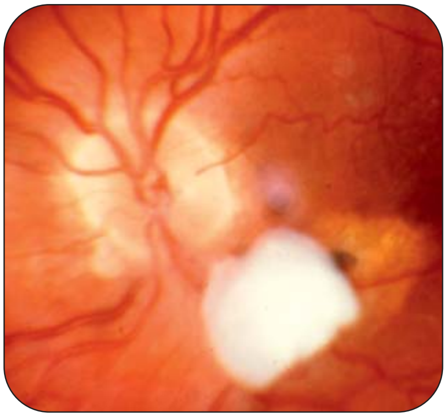

Fig. 12.71

Serpiginous choroidopathy

Definition

- An idiopathic, chronic, recurrent, inflammatory disease which is usually bilateral.

Signs

- Grey-white to yellow lesions that typically start at the edge of the disc and spread outwards (**Fig. 12.72**).

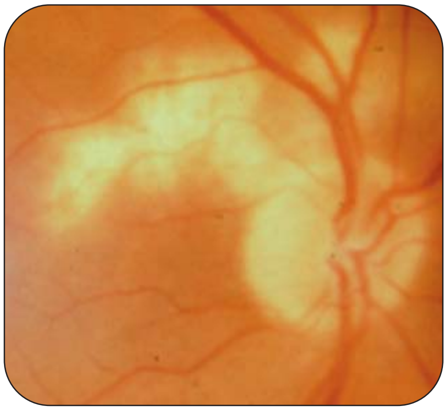

Fig. 12.72

Neoplastic

Choroidal osteoma (osseous choristoma)

Definition

- A rare, benign, slow-growing, ossifying tumour that may involve both eyes.

Signs

- Orange-yellow lesion with scalloped borders which may start at the edge of the disc (**Fig. 12.73**).

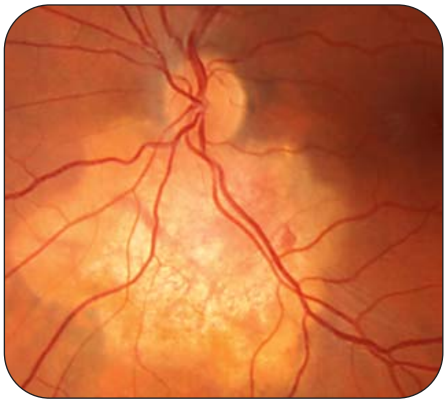

Fig. 12.73

Choroidal melanoma

Signs

- A juxtapapillary location is uncommon (**Fig. 12.74**).

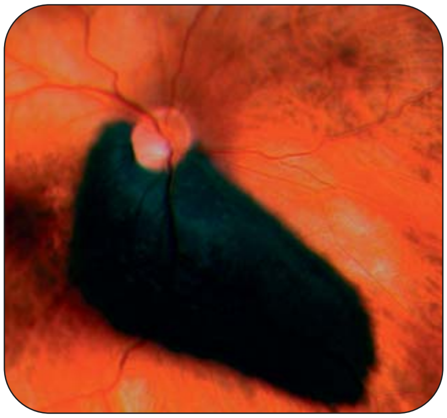

Fig. 12.74

Fundus 13

VASCULAR LESIONS **277**

Haemorrhages **277**
  - *Preretinal (subhyaloid)* 277
  - *Flame-shaped* 277
  - *Roth spots* 278
  - *Intraretinal* 278
  - *Subretinal* 278
  - *Sub-RPE* 279
  - *Choroidal* 279

Cotton-wool spots **280**
  - *Retinal vein occlusion* 280
  - *Preproliferative diabetic retinopathy* 280
  - *Severe hypertensive retinopathy* 281
  - *Purtscher retinopathy* 281
  - *HIV microangiopathy* 281
  - *Radiation retinopathy* 282
  - *Blood dyscrasias* 282
  - *Acute papilloedema* 282
  - *Interferon* 283
  - *Collagen vascular diseases* 283
  - *Miscellaneous causes* 283
  - *Differential diagnosis* 284

Hard exudates arranged in clumps **284**
  - *Background diabetic retinopathy* 285
  - *Old branch retinal vein occlusion* 285
  - *Wet age-related macular degeneration* 285
  - *Retinal artery macroaneurysm* 286
  - *Radiation retinopathy* 286
  - *Retinal telangiectasia* 286
  - *Differential diagnosis* 287

Stellate maculopathy **287**
  - *Severe hypertension* 287
  - *Acute papilloedema* 288
  - *Neuroretinitis* 288
  - *Haemangioblastoma* 288
  - *Cytomegalovirus retinitis* 289

Subretinal hard exudates **289**
  - *Wet age-related macular degeneration* 289
  - *Coats disease* 289

Neovascularisation **290**
  - *Central and peripheral* 290
  - *Peripheral* 291

Retinal emboli **292**
  - *Common* 292
  - *Uncommon* 293

Retinal vascular malformations **294**
  - *Retinal artery macroaneurysm* 294
  - *Coats disease* 294
  - *Congenital retinal macrovessel* 295
  - *Congenital arteriovenous communication* 295

Retinal vascular tumours **295**
  - *Haemangioblastoma (retinal capillary haemangioma)* 295
  - *Retinal racemose haemangioma* 296
  - *Retinal cavernous haemangioma* 296

Changes in calibre **297**

INFLAMMATORY LESIONS **299**

Vasculitis **299**
  - *Periphlebitis* 299
  - *Periarteritis* 300
  - *Frosted branch angiitis* 301

Chorioretinitis with systemic associations **301**
  - *Toxoplasmosis* 301
  - *Toxocariasis* 302
  - *Candidiasis* 302
  - *Sarcoidosis* 302
  - *Behçet disease* 303
  - *Vogt–Koyanagi–Harada syndrome (VKH)* 304
  - *Tuberculous* 305
  - *Acquired syphilis* 305
  - *Choroidal pneumocystosis* 305

Chorioretinitis without systemic associations **306**
  - *Sympathetic ophthalmitis* 306
  - *Multifocal choroiditis with panuveitis syndrome* 306
  - *Presumed ocular histoplasmosis syndrome* 307
  - *Punctate inner choroidopathy* 307
  - *Birdshot retinochoroidopathy* 308
  - *Acute posterior multifocal placoid pigment epitheliopathy* 308
  - *Serpiginous choroidopathy* 309

Necrotising retinitis **309**
  - *Cytomegalovirus retinitis* 309
  - *Acute retinal necrosis* 309
  - *Progressive outer retinal necrosis* 310

MACULAR LESIONS **310**

Oedema **310**
  - *Non-cystoid* 310
  - *Cystoid* 311

Serous detachment **312**
  - *Idiopathic central serous retinopathy* 312
  - *Congenital optic disc pit* 312
  - *Other causes* 313

Detachment of the RPE **313**

Choroidal neovascularisation **314**

Atrophic maculopathy **317**
  - *Dry age-related macular degeneration* 317
  - *Myopic maculopathy* 317
  - *Stargardt macular dystrophy* 317
  - *Central areolar choroidal dystrophy* 318

Bull's eye maculopathy **319**

Fibrotic lesions **320**
  - *Cellophane maculopathy* 320
  - *Pucker* 320
  - *Subretinal fibrosis (disciform scar)* 320
  - *Diffuse subretinal fibrosis* 321

Hereditary crystalline maculopathy **321**
  - *Primary oxalosis type 1* 321
  - *Bietti crystalline dystrophy* 322
  - *AD crystalline retinopathy* 322
  - *Sjögren–Larsson syndrome* 322
  - *Cystinosis* 322

Drug-induced crystalline maculopathy **323**
  - *Tamoxifen* 323
  - *Canthaxanthin* 323
  - *Methoxyflurane* 323

Miscellaneous crystalline maculopathy **324**
  - *Drusen* 324
  - *Gyrate atrophy* 324

2A bilateral idiopathic acquired parafoveolar telangiectasia 324

Macular hole 325
- *Idiopathic (age-related)* 325
- *Microhole* 325
- *Myopic* 326
- *Traumatic* 326
- *Causes of macular pseudo-hole* 326

PERIPHERAL RETINAL DEGENERATIONS 327

Innocuous 327
- *Honeycomb (reticular)* 327
- *Peripheral drusen* 327
- *Microcystoid* 327
- *Pavingstone* 328

Predisposing 328
- *Lattice* 328
- *Snailtrack* 328
- *White-without-pressure* 328

RETINAL DETACHMENT 329

Rhegmatogenous 329
- *Fresh* 329
- *Proliferative vitreoretinopathy* 330
- *Long-standing* 330

Tractional 330

Exudative 331

Differential diagnosis 331
- *Degenerative retinoschisis* 331
- *Choroidal detachment* 332
- *Uveal effusion syndrome* 332

SCATTERED PIGMENTARY CHANGES 332

Clumping 332
- *Chloroquine retinotoxicity* 332
- *Thioridazine retinotoxicity* 333
- *‘Leopard spots’* 333
- *Grouped congenital hypertrophy of the RPE* 333
- *Retinitis pigmentosa* 334
- *End-stage acquired syphilitic chorioretinitis* 334
- *Pigmented paravenous chorioretinal atrophy* 334
- *Enhanced S-cone syndrome* 335

Mottling 335
- *Rubella retinopathy* 335
- *Congenital syphilis* 335
- *‘Peau d’orange’* 336
- *Female carriers of choroideremia* 336

MULTIPLE YELLOW-WHITE SPOTS AND FLECKS 336
- *Hard drusen* 336
- *Soft drusen* 337
- *Cuticular (basal laminar) drusen* 337
- *Familial drusen (Doyne honeycomb dystrophy)* 337
- *Malattia levantinese* 338
- *Fundus flavimaculatus* 338
- *Fundus albipunctatus* 338
- *Retinitis punctata albescens* 339
- *Benign flecked retina* 339
- *Alport syndrome* 339
- *North Carolina macular dystrophy* 340
- *Sorsby pseudo-inflammatory dystrophy* 340

FLAT FOCAL LESIONS 340

Black 340
- *Typical congenital RPE hypertrophy* 340
- *Atypical congenital RPE hypertrophy* 341
- *Choroidal naevus* 341
- *RPE hamartoma* 341
- *Bilateral diffuse uveal melanocytic proliferation* 342

Pale 342
- *Old retinochoroiditis* 342
- *Idiopathic chorioretinal atrophy* 342
- *Congenital hypertrophy of the RPE* 343
- *Coloboma of the retinal and choroid* 343
- *Congenital albinotic spots* 343

Egg-yolk 344
- *Best vitelliform macular dystrophy* 344
- *Adult vitelliform macular dystrophy* 344
- *Multifocal vitelliform lesions* 344

Cherry-red spot 345
- *Acute central retinal artery occlusion* 345
- *Sphingolipidoses* 345

FOCAL ELEVATED LESIONS 345

Retinal 346
- *Retinal astrocytoma* 346
- *Retinoblastoma* 346
- *Retinoma* 346
- *Toxocara granuloma* 347

Choroidal 347
- *Choroidal melanoma* 347
- *Circumscribed choroidal haemangioma* 348
- *Choroidal metastatic carcinoma* 349
- *Choroidal granuloma* 349

DIFFUSE CHORIORETINAL ATROPHY 350

Hereditary 350
- *Choroideremia* 350
- *Generalised choroidal dystrophy* 350
- *Gyrate atrophy* 350
- *Progressive bifocal chorioretinal atrophy* 351

Acquired 351
- *High myopia* 351
- *Drug-induced* 351
- *Serpiginous chorioretinopathy* 352
- *Siderosis retinopathy* 352
- *Onchocercal chorioretinitis* 352

CHORIORETINAL FOLDS 353

DIFFERENTIAL DIAGNOSIS OF VISUAL SYMPTOMS 354

VASCULAR LESIONS

Haemorrhages

Preretinal (subhyaloid)

Location

- The haemorrhage originates from the superficial retinal vessels and is located between the retina and posterior vitreous face.

Signs

- Usually solitary, round and frequently located at the posterior pole, where it obscures all underlying retinal landmarks (**Fig. 13.1**).
- With time the blood may settle with gravity, giving rise to the typical boat-shaped crescentic configuration (**Fig. 13.2**).
- In some cases, a large haemorrhage may break through into the vitreous cavity.

Causes

- Proliferative retinopathies (e.g. diabetic, following venous occlusion, radiation, sickle cell, ocular ischaemic syndrome, retinopathy of prematurity and vasculitis).
- Retinal artery macroaneurysm.
- Trauma.
- Subarachnoid haemorrhage (Terson syndrome).
- Valsalva retinopathy caused by a severe increase in venous pressure brought on by strenuous physical exertion.
- Non-accidental injury (‘shaken-baby’ syndrome).

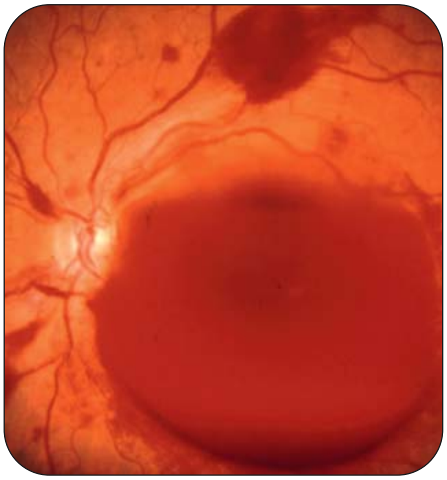

Fig. 13.1

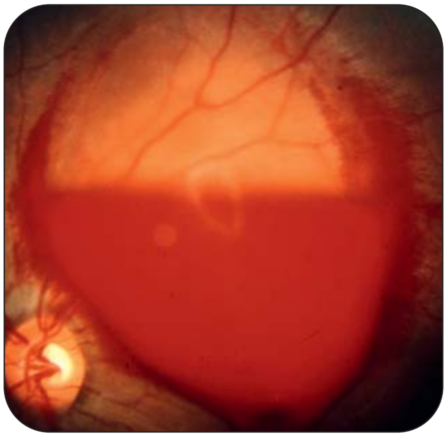

Fig. 13.2

Flame-shaped

Location

- The haemorrhage originates from the superficial pre-capillary arterioles and is located within the retinal nerve fibre layer.

Signs

- Bright red, usually multiple haemorrhages spreading along the nerve fibre layer (**Fig. 13.3**).

Causes

- Retinal vein occlusion.
- Hypertensive retinopathy.
- Background diabetic retinopathy.
- Retinal periphlebitis.
- Blood dyscrasias.

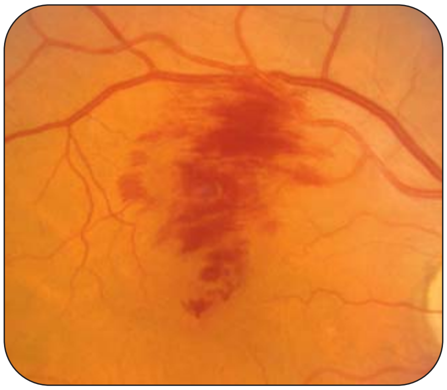

Fig. 13.3

- Optic nerve disease – acute papilloedema, anterior ischaemic optic neuropathy and glaucoma.

Roth spots

Signs

- Flame-shaped haemorrhages with white centres (**Fig. 13.4**).

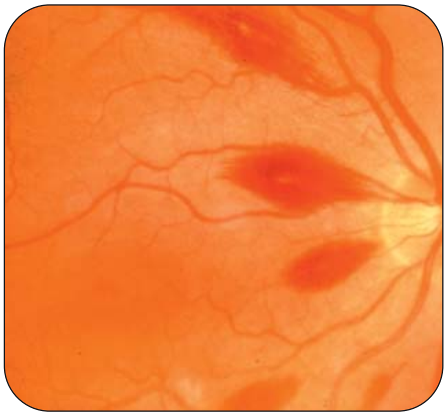

Fig. 13.4

Causes

- Severe anaemia.
- Leukaemia.
- Dysproteinaemia.
- HIV microangiopathy.
- Bacterial endocarditis.

Intraretinal

Location

- The haemorrhage originates from the venous end of the deep retinal capillaries and is located within the sensory retina.

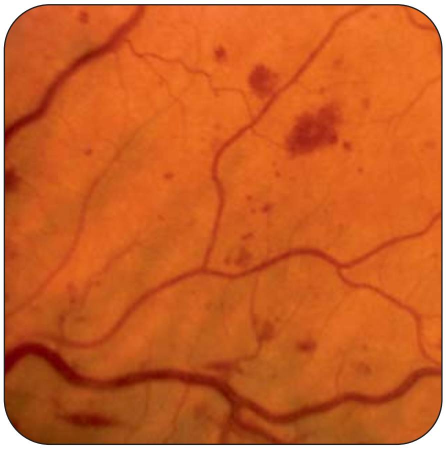

Fig. 13.5

Signs (Fig. 13.5)

a. *Dot* haemorrhages are small and round, and located in the compact middle layers of the retina; they may be difficult to distinguish from microaneurysms.

b. *Blot* haemorrhages occupy the full thickness of the retina and are larger, darker and deeper than dot haemorrhages.

c. *Large blotchy* haemorrhages are usually indicative of retinal ischaemia.

Causes

- Background diabetic retinopathy.
- Retinal vein occlusion.
- Radiation retinopathy.

Subretinal

Location

- The haemorrhage is located between the photoreceptors and the retinal pigment epithelium (RPE).

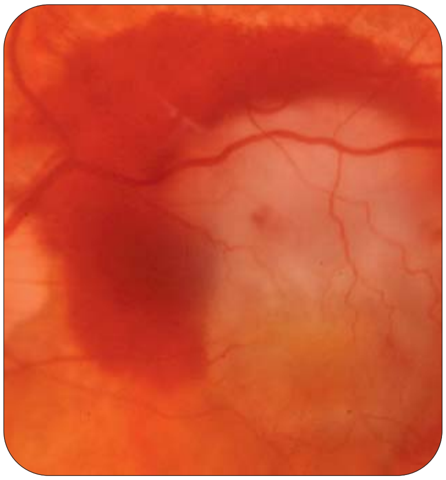

Fig. 13.6

Signs

- Usually large and bright red with slightly indistinct borders (**Fig. 13.6**).
- The retina overlying the haemorrhage is usually slightly elevated, but the retinal vessels are clearly seen.

Causes

- Choroidal neovascularisation (CNV).
- Ruptured retinal artery macroaneurysm.
- Coats disease.
- Sickle cell anaemia.
- Blunt ocular trauma.

Sub-RPE

Location

- The haemorrhage is located between the RPE and Bruch membrane.

Signs

- Slightly elevated, very dark-red, almost black, with relatively well-defined borders.
- They are often solitary and located at the macula (**Fig. 13.7**).

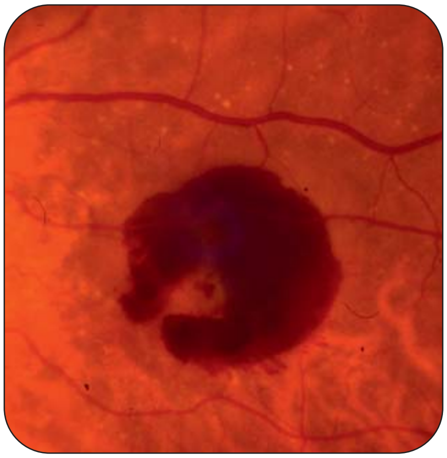

Fig. 13.7

Causes

- CNV is by far the most common (haemorrhagic RPE detachment).

Choroidal

Signs

- Dark red and frequently extensive.

Causes

- Blunt trauma.
- Complication of drainage of subretinal fluid during retinal detachment surgery (**Fig. 13.8**).

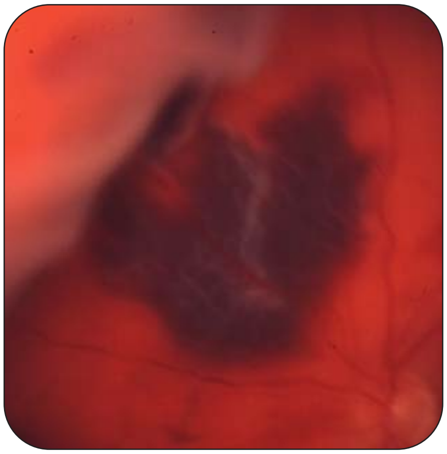

Fig. 13.8

Cotton-wool spots

Cotton-wool spots are accumulations of neuronal debris that results from disruption of nerve axons. They appear as small, whitish, fluffy superficial lesions in the post-equatorial fundus (**Fig. 13.9**). The causes are as follows:

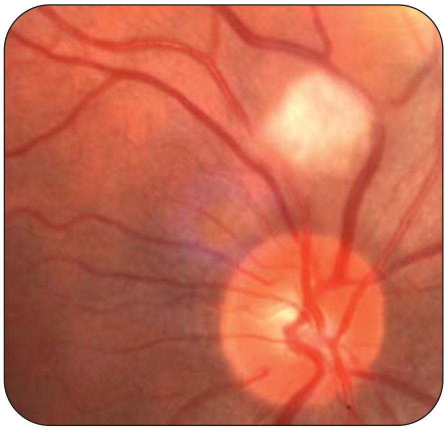
Fig. 13.9

Retinal vein occlusion

Signs

- Cotton-wool spots, venous dilatation, and flame-shaped and blotchy haemorrhages (**Fig. 13.10A**).
- FA shows venous staining, capillary non-perfusion, blockage by blood and pruning of vessels in the affected area (**Fig. 13.10B**).

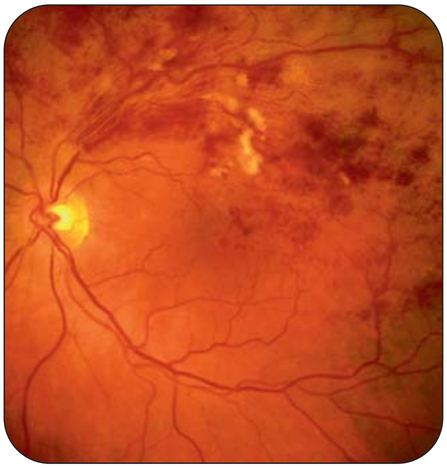
Fig. 13.10A

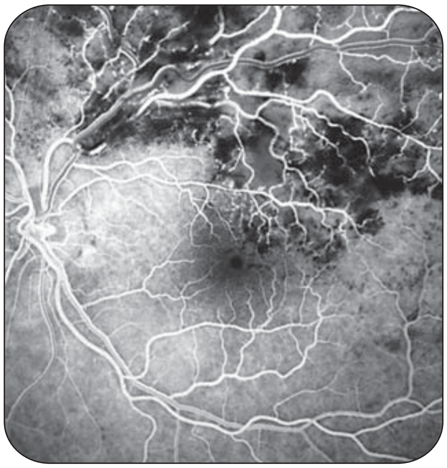
Fig. 13.10B

Preproliferative diabetic retinopathy

Signs

- Cotton-wool spots, intraretinal microvascular abnormalities (IRMA), venous dilatation and beading, and haemorrhages (**Fig. 13.11A**).
- Peripheral arteriolar narrowing, silver-wiring and occlusion.
- FA shows extensive capillary non-perfusion, punctate staining of microaneurysms and enhancement of the venous changes (**Fig. 13.11B**).

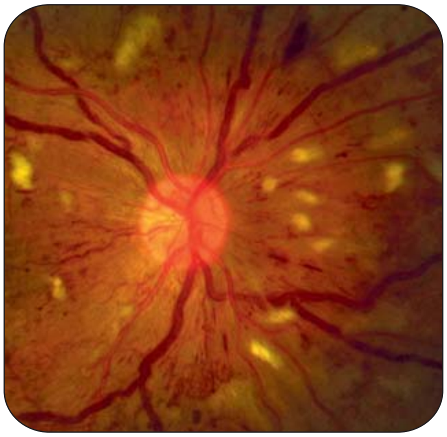
Fig. 13.11A

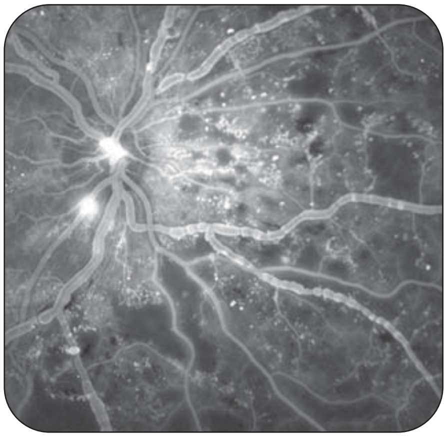
Fig. 13.11B

Severe hypertensive retinopathy

Signs

- Cotton-wool spots, arteriolar narrowing, arteriolosclerosis, flame-shaped haemorrhages, retinal oedema, macular star and disc oedema (**Fig. 13.12**).

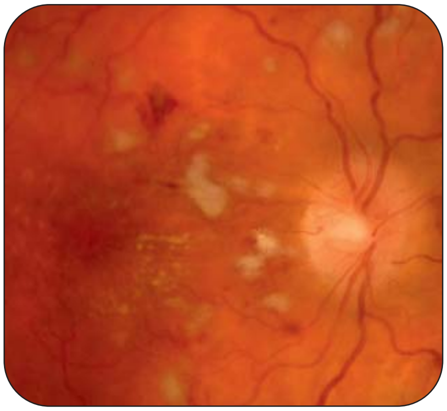

Fig. 13.12

Purtscher retinopathy

Definition

- A rare condition that has many causes (Table 13.1).

Signs

- Large cotton-wool spots, some of which may be confluent (**Fig. 13.13**).
- Superficial peripapillary haemorrhages are common.

Table 13.1 **Causes of Purtscher retinopathy**

1. **Severe trauma**
   - Head
   - Chest
   - Long bones
2. **Embolism**
   - Fat
   - Air
   - Amniotic fluid
3. **Pancreatic disease**
   - Acute pancreatitis
   - Carcinoma
4. **Miscellaneous**
   - Thrombocytopenic purpura
   - Lymphoma
   - Bone marrow transplantation
   - Chronic renal failure

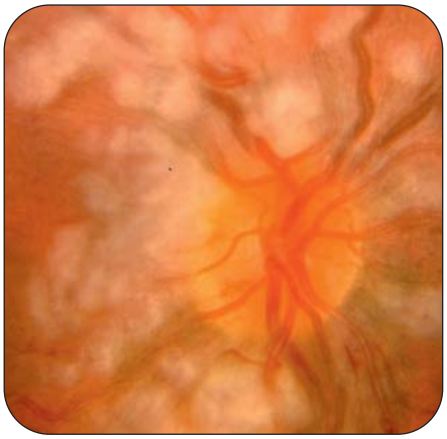

Fig. 13.13

HIV microangiopathy

Signs

- Cotton-wool spots (**Fig. 13.14**) occasionally associated with retinal haemorrhages and capillary abnormalities.

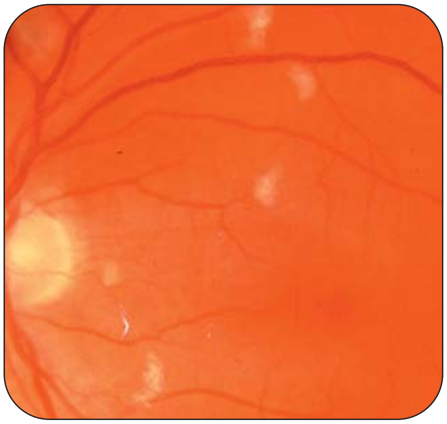

Fig. 13.14

Radiation retinopathy

Signs

- Cotton-wool spots, haemorrhages (**Fig. 13.15**), microaneurysms, macular oedema, hard exudates, proliferative retinopathy and papillopathy.

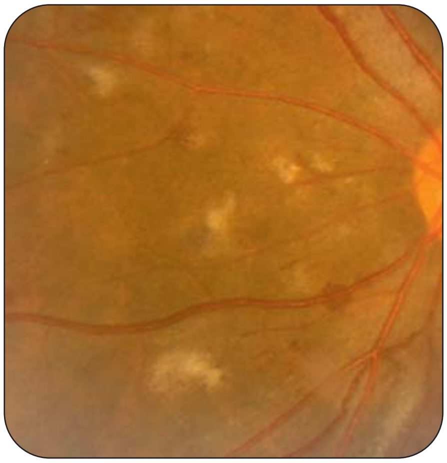

Fig. 13.15

Blood dyscrasias

Signs

- Cotton-wool spots, venous tortuosity, dot-blot and flame-shaped retinal haemorrhages and Roth spots (**Fig. 13.16**).

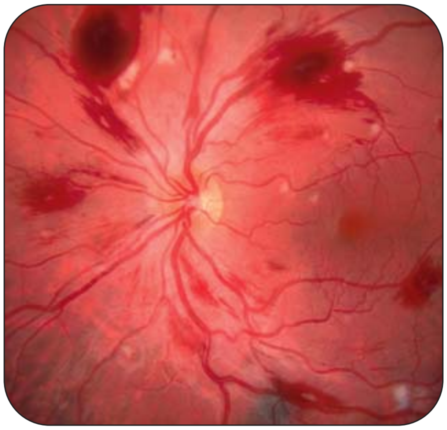

Fig. 13.16

Acute papilloedema

Signs

- Bilateral peripapillary cotton-wool spots and disc swelling (**Fig. 13.17**).

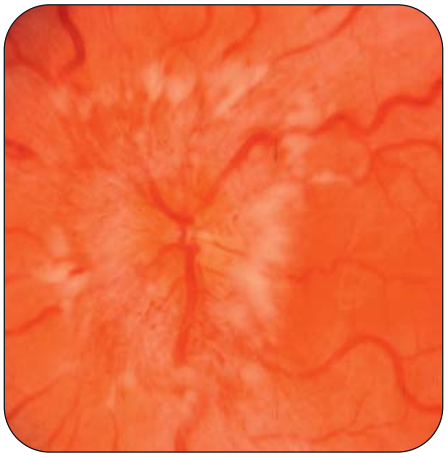

Fig. 13.17

Interferon

Interferon administration particularly with high dosage may cause cotton-wool spots (**Fig. 13.18**).

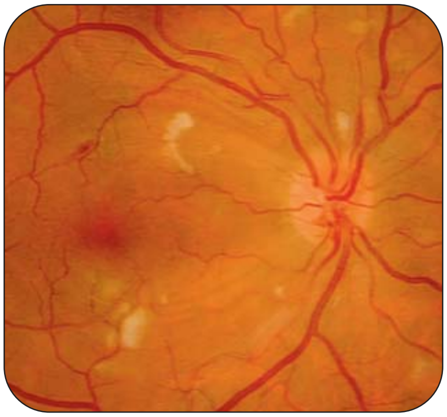

Fig. 13.18

Collagen vascular diseases

a. *Scleroderma* – fixed facial expression, facial rash and 'beaking' of the nose (**Fig. 13.19**).

b. *Systemic lupus erythematosus* – 'butterfly' skin rash (**Fig. 13.20**).

c. *Dermatomyositis* – heliotrope facial rash and focal ulceration (**Fig. 13.21**).

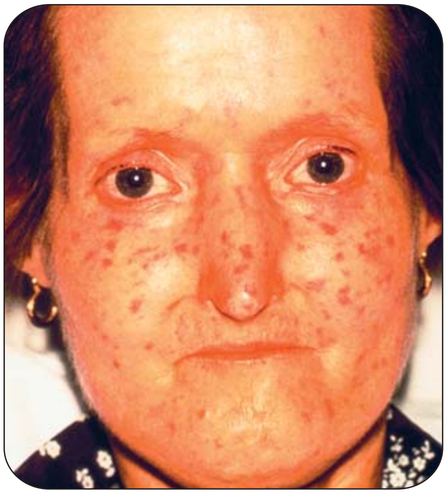

Fig. 13.19

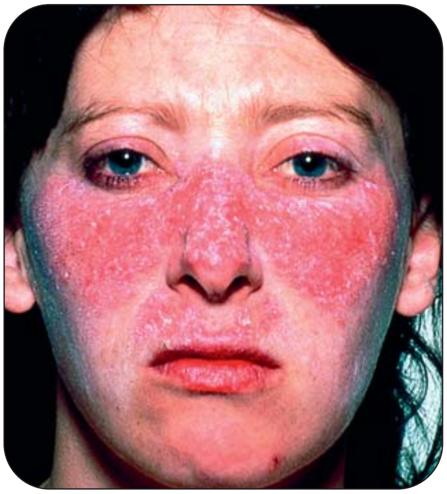

Fig. 13.20

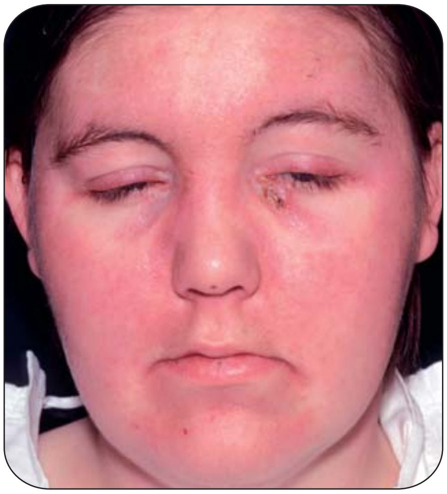

Fig. 13.21

Miscellaneous causes

a. *Ocular ischaemic syndrome.*

b. *Goodpasture syndrome.*

c. *Antiphospholipid antibody syndrome.*

Differential diagnosis

- Congenital myelinated nerve fibres – white feathery streaks within the nerve fibre layer (**Fig. 13.22**).
- Retinitis – superficial yellow-white cloudy lesions associated with vitritis (**Fig. 13.23**).

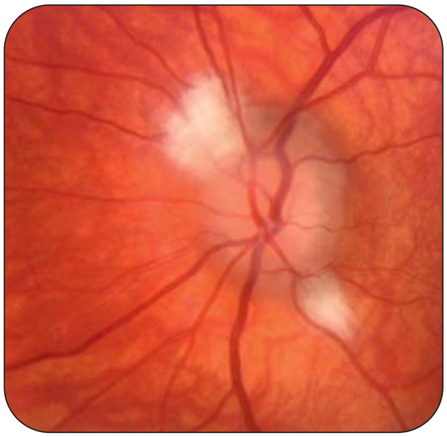

Fig. 13.22

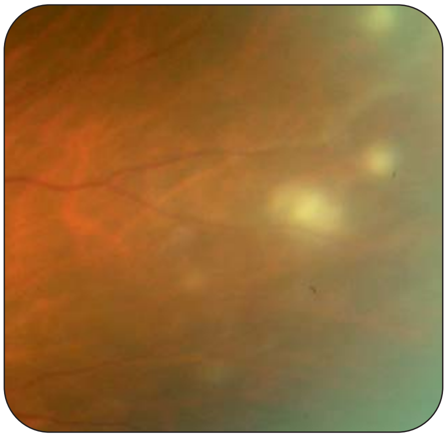

Fig. 13.23

Hard exudates arranged in clumps

Retinal hard exudate formation is caused by chronic capillary leakage. The exudates develop at the junction of normal and oedematous retina and are composed of lipoprotein and lipid-filled macrophages. They are located mainly within the outer plexiform layer. Clinically they appear as waxy yellow plaques with relatively distinct margins, most frequently located at the posterior pole (**Fig. 13.24**). The causes are as follows:

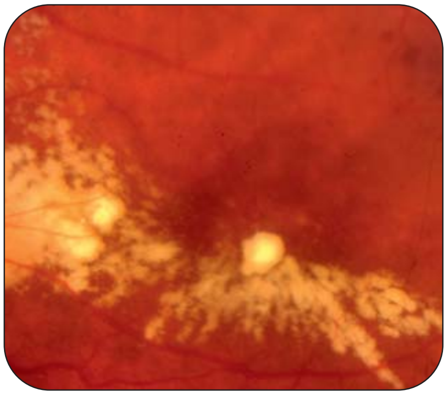

Fig. 13.24

Background diabetic retinopathy

Signs

- Bilateral, hard exudates, microaneurysms and retinal haemorrhages (**Fig. 13.25**).

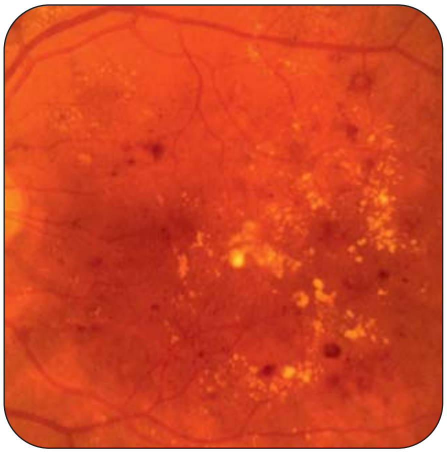

Fig. 13.25

Old branch retinal vein occlusion

Signs

- Unilateral segmental hard exudates, venous sheathing, collaterals and residual haemorrhage (**Fig. 13.26**).

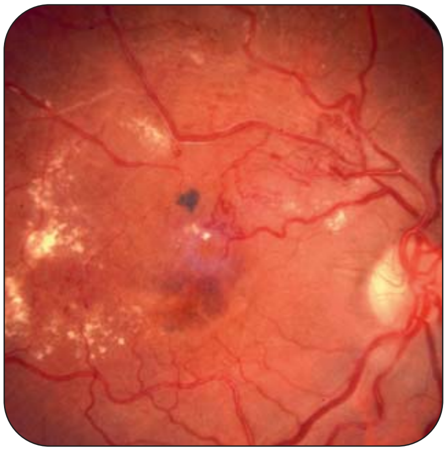

Fig. 13.26

Wet age-related macular degeneration

Signs

- Small hard exudates**,** soft drusen and macular haemorrhage (**Fig. 13.27**).

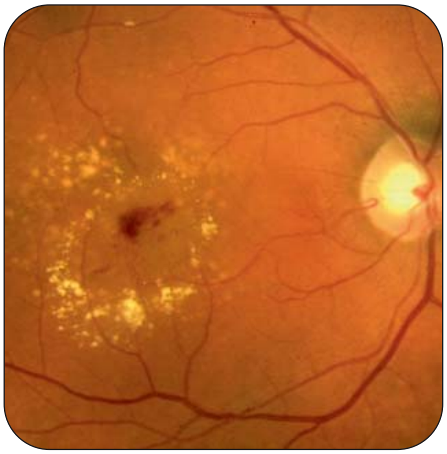

Fig. 13.27

Retinal artery macroaneurysm

Signs

- Unilateral ring of hard exudates surrounding the lesion (**Fig. 13.28**).
- Associated haemorrhages may be subretinal, intraretinal, preretinal and vitreous.

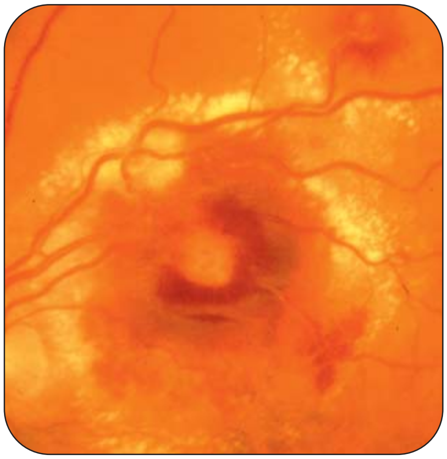
Fig. 13.28

Radiation retinopathy

- Unilateral hard exudates, microaneurysms and macular oedema (**Fig. 13.29**).
- Cotton-wool spots, proliferative retinopathy and papillopathy.

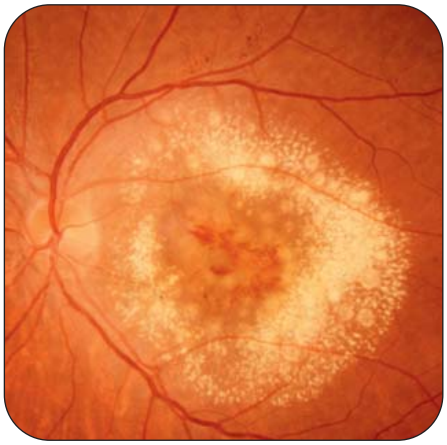
Fig. 13.29

Retinal telangiectasia

Signs

- Intraretinal and subretinal exudates, retinal vascular tortuosity, dilatation and aneurysm formation (**Fig. 13.30**).

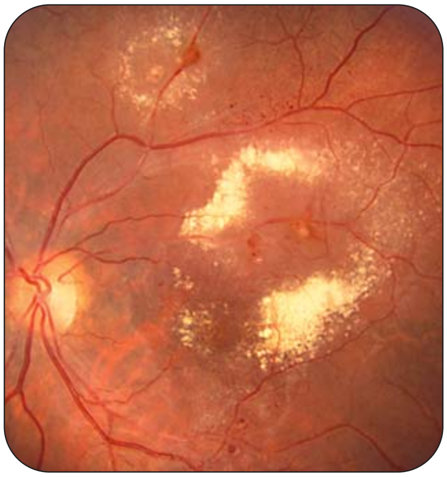
Fig. 13.30

Differential diagnosis

- Macular drusen are bilateral, focal yellow spots which are not arranged in clumps or rings and are not associated with retinal microvascular changes (**Fig. 13.31**).

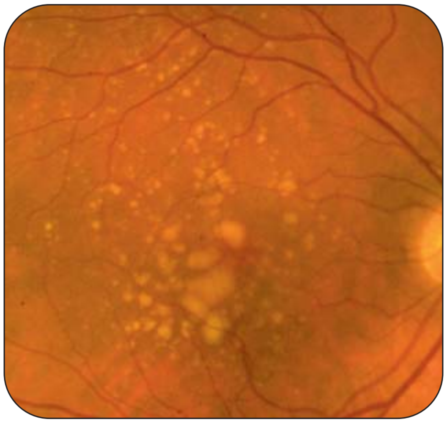

Fig. 13.31

Stellate maculopathy

In stellate maculopathy the retinal exudates develop within the radially arranged nerve fibre layer of Henle (**Fig. 13.32**) and are frequently associated with disc swelling. The causes are as follows:

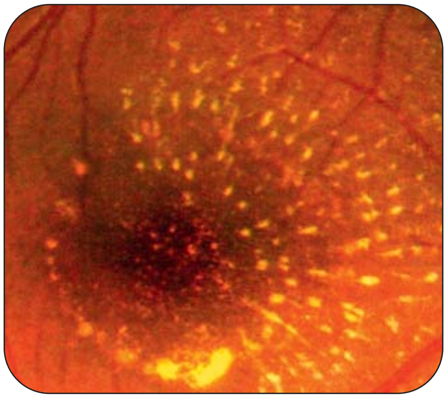

Fig. 13.32

Severe hypertension

Signs

- Cotton-wool spots, flame-shaped haemorrhages, arteriolosclerosis and disc swelling in the malignant (accelerated) phase (**Fig. 13.33**).

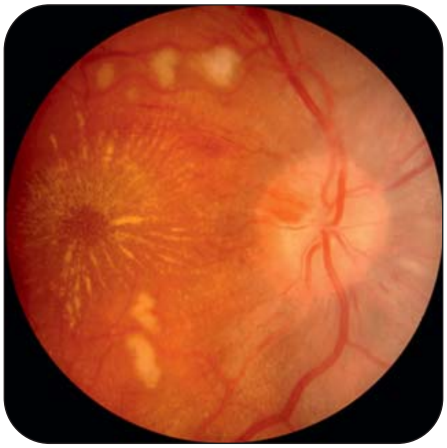

Fig. 13.33

Acute papilloedema

Signs

- Severe bilateral disc swelling (**Fig. 13.34**).

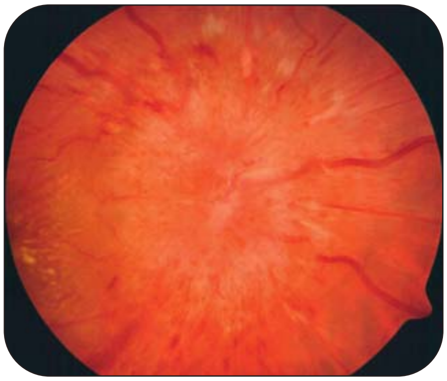

Fig. 13.34

Neuroretinitis

Signs

- Mild disc swelling which is subsiding as the macular star forms (**Fig. 13.35**).

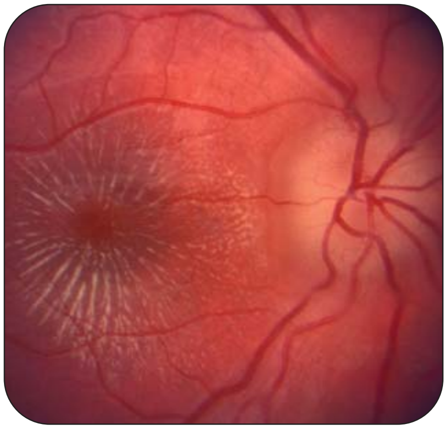

Fig. 13.35

Haemangioblastoma

Signs

- Stellate maculopathy may occur with a lesion on the disc (**Fig. 13.36**) or in the periphery.

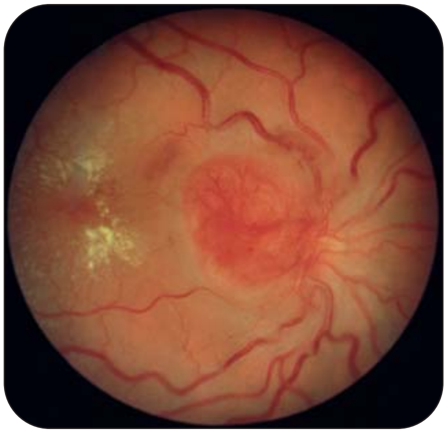

Fig. 13.36

Cytomegalovirus retinitis

Signs

- Retinal opacification along the vascular arcades and haemorrhages (**Fig. 13.37**).

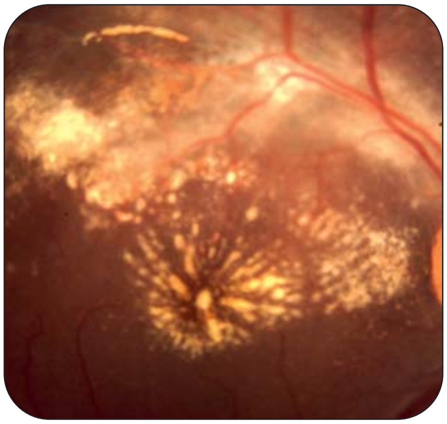

Fig. 13.37

Subretinal hard exudates

Subretinal exudates are caused by massive chronic leakage from CNV or other subretinal vascular lesions and are frequently associated with exudative RD. Important causes are:

Wet age-related macular degeneration

Signs

- Extensive subretinal exudation which may be unilateral or bilateral (**Fig. 13.38**).

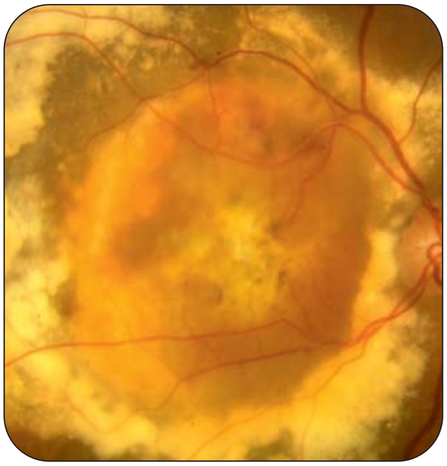

Fig. 13.38

Coats disease

Signs

- Extensive subretinal exudation associated with retinal telangiectasia (see Fig. 13.30).

Neovascularisation

Neovascularisation is caused by angiogenic growth factors elaborated by hypoxic retinal tissues in an attempt to revascularise affected retina. The new vessels may be located in the central fundus or in the periphery.

Central and peripheral

Signs

- New vessels may involve the optic nerve head (new disc vessels – NDV – **Fig. 13.39**) or develop along the major vascular arcades (new vessels elsewhere – NVE – **Fig. 13.40**).
- The vessels may be flat or elevated, and they may be bare or associated with fibrosis (**Fig. 13.41**).
- FA shows intensive leakage of dye and capillary non-perfusion (**Fig. 13.42**).

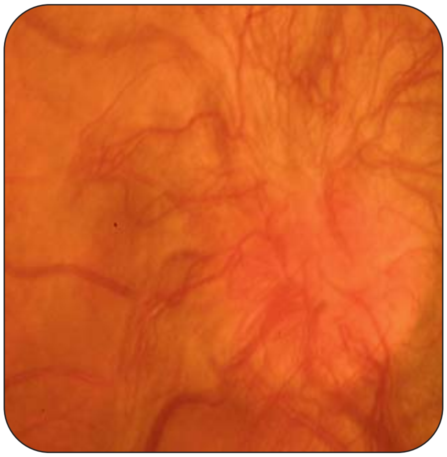
Fig. 13.39

Causes

- Proliferative diabetic retinopathy (**Fig. 13.43**).
- Old branch retinal vein occlusion (**Fig. 13.44**).
- Ocular ischaemic syndrome.
- Radiation retinopathy.
- Retinal vasculitis (**Fig. 13.45**).

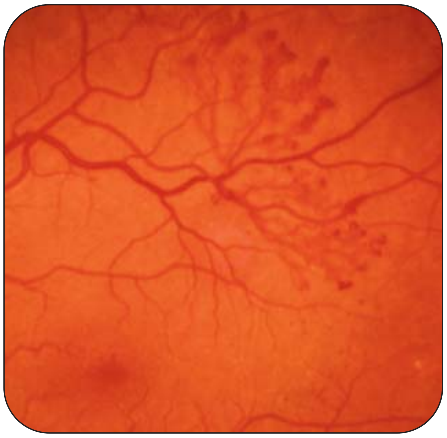
Fig. 13.40

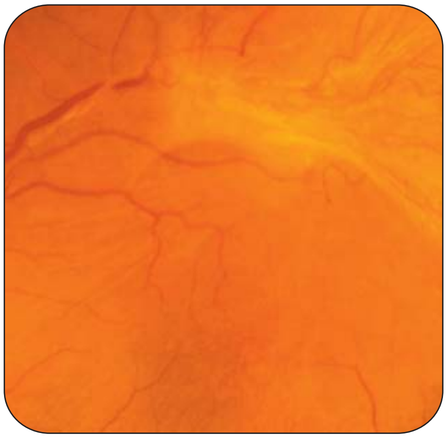
Fig. 13.41

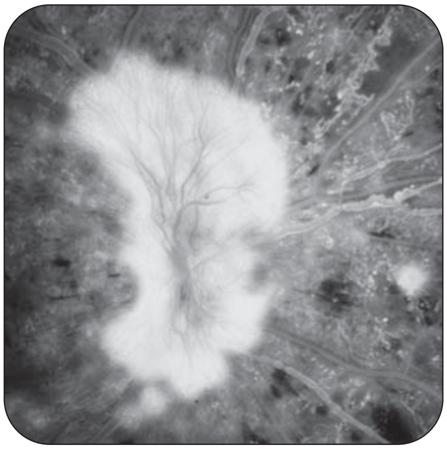
Fig. 13.42

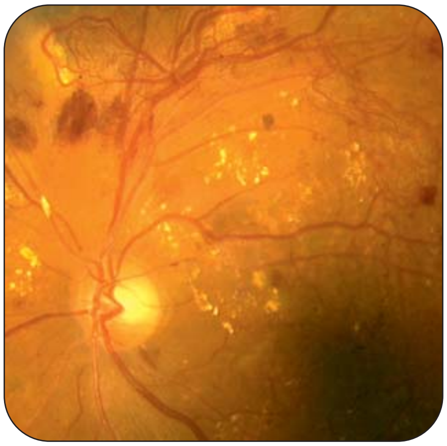
Fig. 13.43

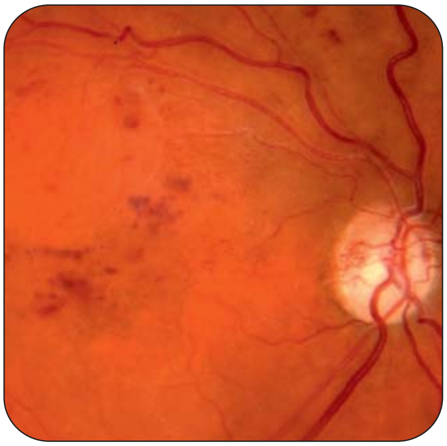
Fig. 13.44

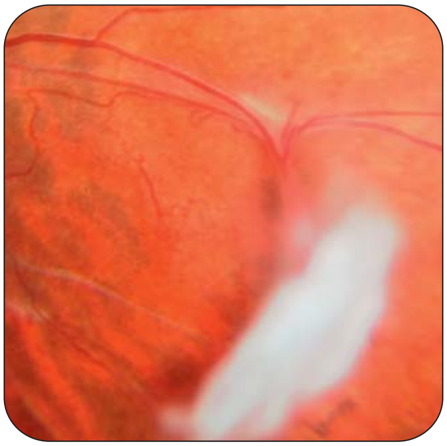
Fig. 13.45

- Previous retinal artery occlusion is an uncommon cause.
- Carotid–cavernous fistula.

Complications

- Vitreous haemorrhage that may be subhyaloid (**Fig. 13.46** and see Figs 13.1 and 13.2) or intragel (**Fig. 13.47** and **Fig. 13.48**).
- Tractional RD (**Fig. 13.49**).

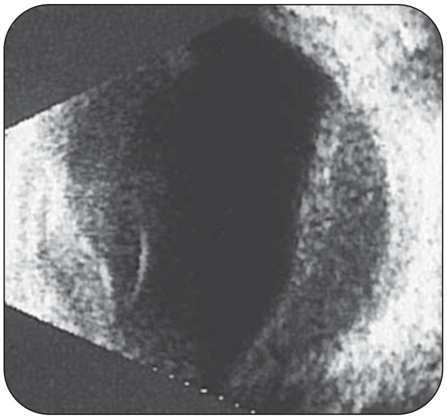
Fig. 13.46

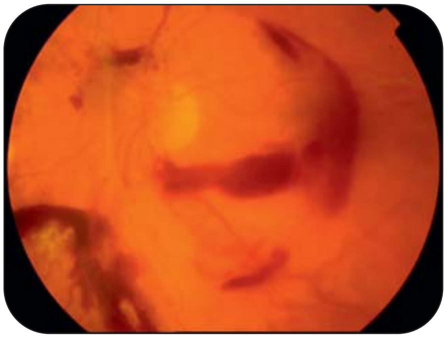
Fig. 13.47

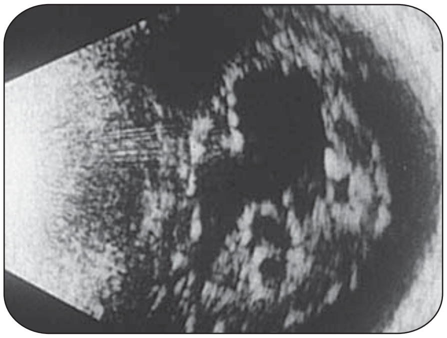
Fig. 13.48

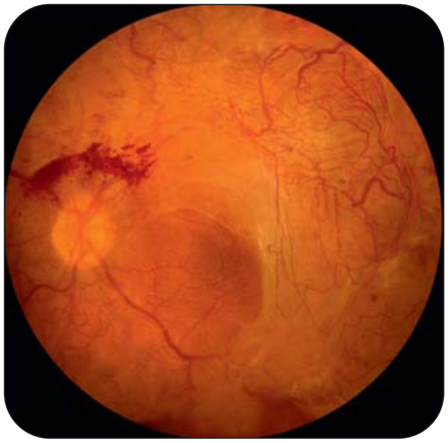
Fig. 13.49

Peripheral

Signs

- New vessels at or anterior to the equator (**Fig. 13.50**).
- They are usually located at the junction of perfused and non-perfused retina.

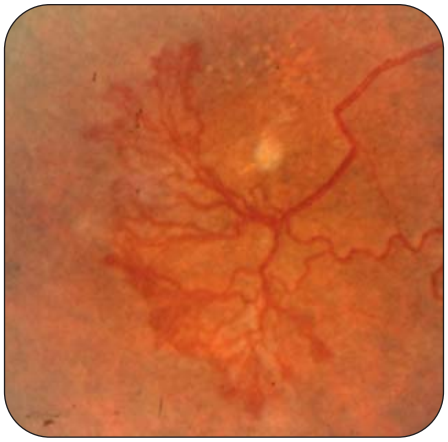
Fig. 13.50

Causes

a. *Sickle cell retinopathy* has a ‘sea-fan’ configuration (**Fig. 13.51**).

b. *Eales disease* – recurrent vitreous haemorrhage from peripheral new vessels (**Fig. 13.52**).

c. *Retinopathy of prematurity* (**Fig. 13.53**).

d. *Familial exudative vitreoretinopathy* (**Fig. 13.54**).

e. *Chronic myeloid leukaemia.*

f. *Sarcoidosis.*

g. *Incontinentia pigmenti* (Bloch–Sulzberger syndrome) – characteristic cutaneous lesions (see Figs 9.21 and 9.22), retinal dysplasia and leukocoria.

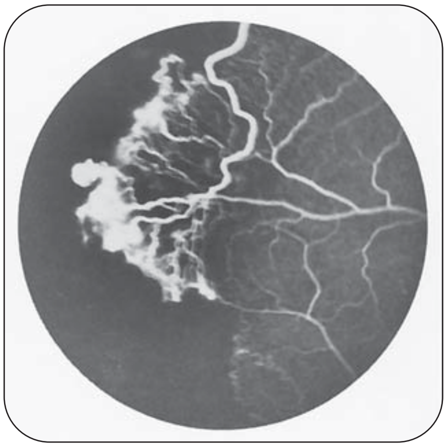

Fig. 13.51

Complications

- Vitreous haemorrhage.
- Tractional retinal detachment.

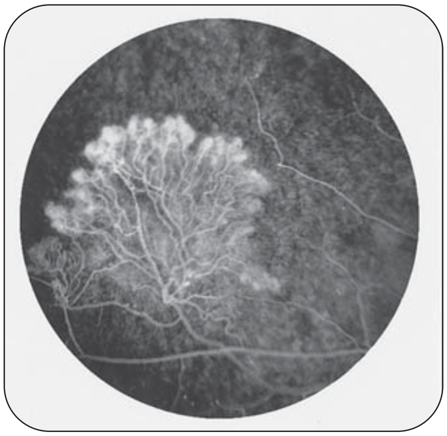

Fig. 13.52

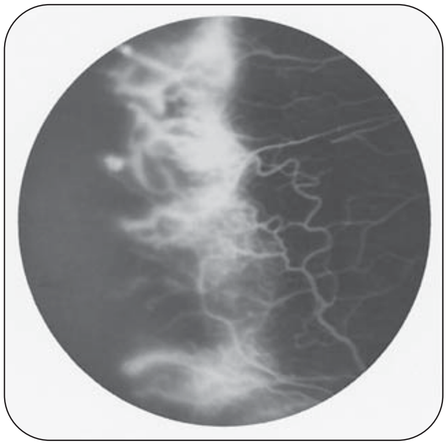

Fig. 13.53

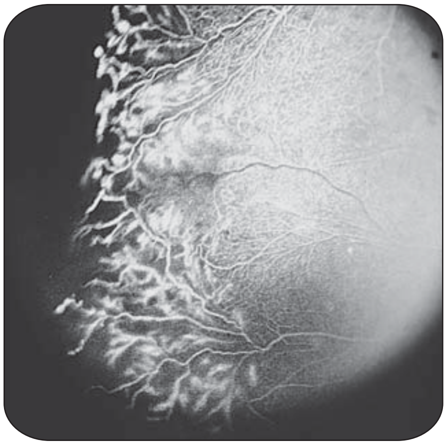

Fig. 13.54

Retinal emboli

Common

a. *Fibrinoplatelet* emboli are dull grey, elongated particles which are usually multiple (**Fig. 13.55**) and may occasionally fill the entire lumen of an arteriole (**Fig. 13.56**).

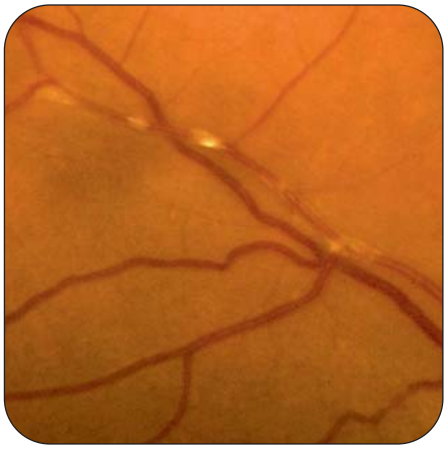

Fig. 13.55

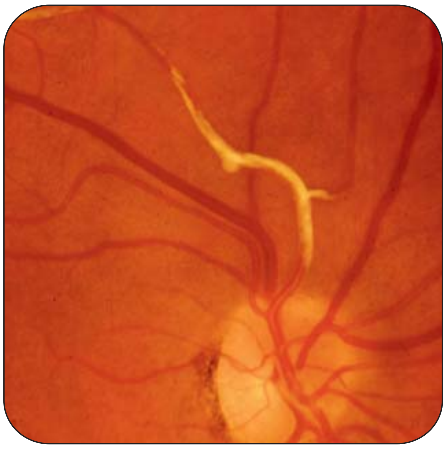

Fig. 13.56

b. *Cholesterol (Hollenhorst) plaques* are often associated with carotid stenosis (**Fig. 13.57**). They are small, glistening, usually multiple and typically located at arteriolar bifurcations (**Fig. 13.58**).

c. *Calcific* emboli originate from cardiac valve disease. They are white, usually solitary and commonly located on the disc (**Fig. 13.59**). They may give rise to major arterial occlusion (**Fig. 13.60**).

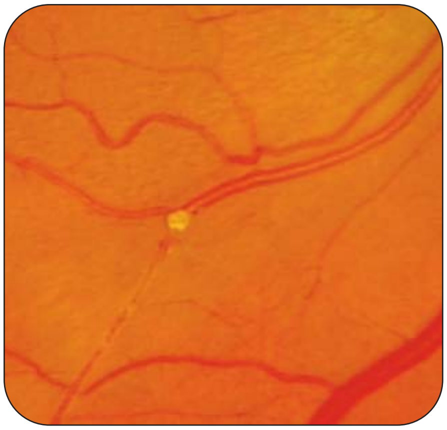

Fig. 13.57

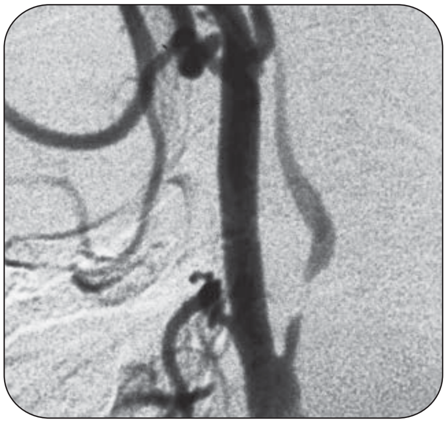

Fig. 13.58

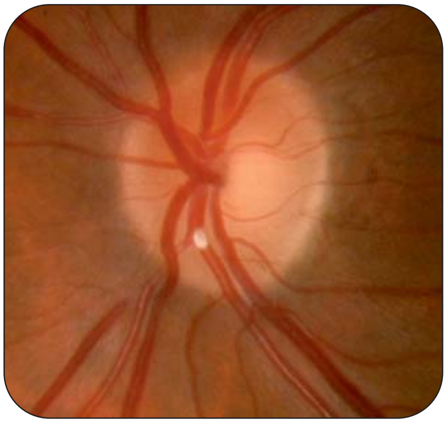

Fig. 13.59

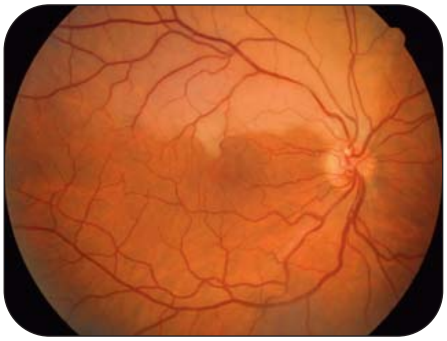

Fig. 13.60

Uncommon

a. *Heart valve vegetations* in bacterial endocarditis.

b. *Myxomatous material* from an atrial myxoma.

c. *Fat emboli seen in trauma* to the chest and long bones (Purtscher retinopathy).

d. *Fat emboli in acute pancreatitis.*

e. *Talc emboli* in intravenous drug abusers.

f. *Metastatic tumour.*

g. *Amniotic fluid.*

Retinal vascular malformations

Retinal artery macroaneurysm

Definition

- An uncommon, usually unilateral condition which typically affects elderly hypertensive females.

Signs

- Localised, fusiform or saccular dilatation usually arising within the first three orders of bifurcation (**Fig. 13.61**).
- More than one lesion in the same eye occurs in about 20% of cases.

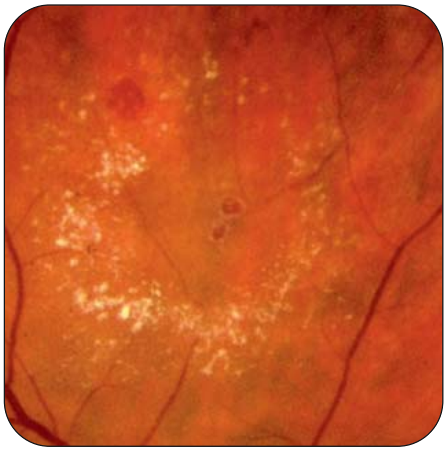
Fig. 13.61

Complications

- Haemorrhage with surrounding hard exudates are common (see Fig. 13.28).

Coats disease

Definition

- An uncommon, developmental vascular anomaly which typically presents at about the age of 8 years with leukocoria (see Fig. 9.34) or strabismus.
- It is more common in boys than in girls.

Signs

- Vascular dilatation, tortuosity and aneurysm formation at the posterior pole and periphery (**Fig. 13.62**).
- Retinal and subretinal exudate formation (see Fig. 13.30).

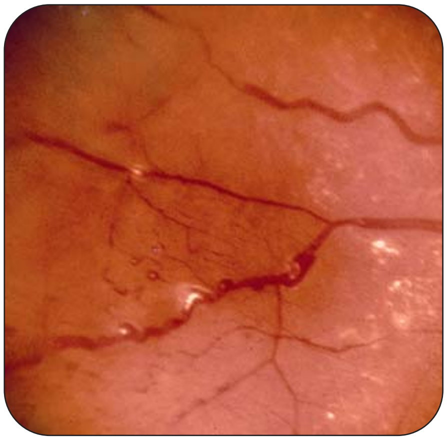
Fig. 13.62

Complications

- Exudative RD and a retrolental mass which may give rise to leukocoria.

Differential diagnosis

- Late-onset retinoblastoma.
- *Toxocara canis* which may also be associated with hard exudate formation.

Congenital retinal macrovessel

Signs

- Unilateral, large, aberrant vessel, usually a vein, in the posterior pole that may cross the fovea and horizontal raphe (**Fig. 13.63**).

Differential diagnosis

- Retinal racemose haemangioma (see Fig. 13.66).

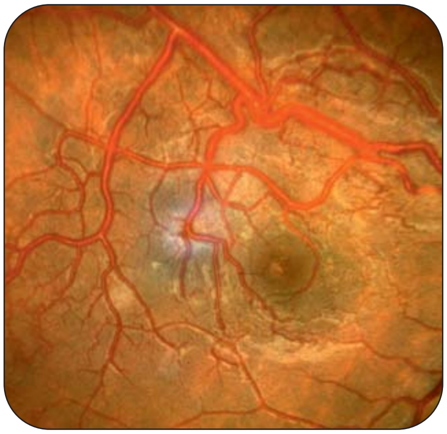

Fig. 13.63

Congenital arteriovenous communication

Signs

- Unilateral anomaly in which both the vein and artery are dilated (**Fig. 13.64**).
- Location is in the papillomacular bundle or the superotemporal quadrant.

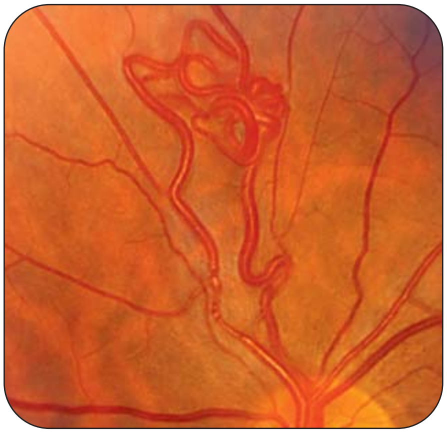

Fig. 13.64

Retinal vascular tumours

Haemangioblastoma (capillary haemangioma)

Definition

- A benign but vision-threatening tumour that may be solitary or multifocal.
- Both eyes are affected in 50% of cases.

Signs

- Round and orange-red tumour.
- Associated with dilated and tortuous artery and vein (**Fig. 13.65**).

Complications

- Hard exudates near the lesion or at the macula.
- Vitreous haemorrhage.
- Retinal detachment.

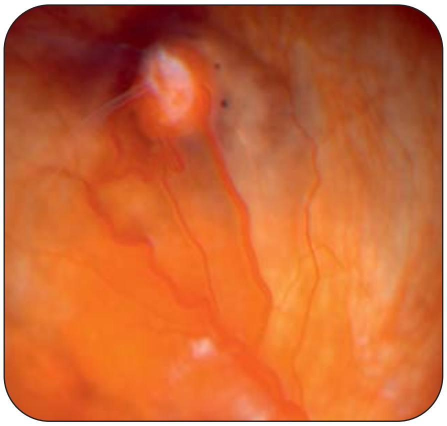

Fig. 13.65

Look for

- Von Hippel–Lindau syndrome (see Fig. 12.48).

Retinal racemose haemangioma

Definition

- A usually unilateral, congenital arteriovenous malformation that may occur in isolation or in association with systemic lesions (Wyburn-Mason syndrome).

Signs

- Grossly dilated and tortuous arteries and veins which are more numerous than in a normal fundus (**Fig. 13.66**).

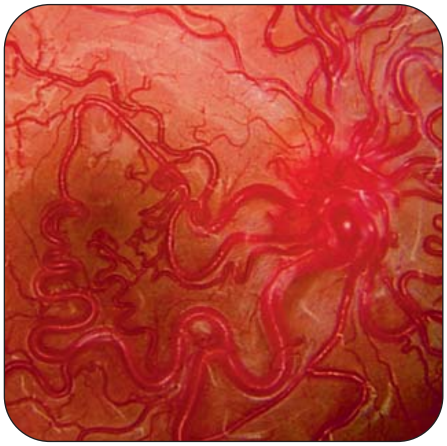

Fig. 13.66

Retinal cavernous haemangioma

Definition

- A congenital, innocuous unilateral tumour.

Signs

- Clumps of thin-walled, grape-like saccular aneurysms filled with blood (**Fig. 13.67**).

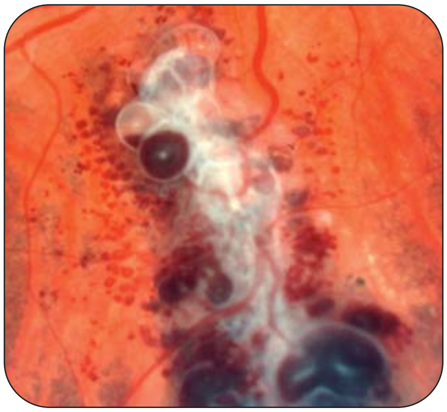

Fig. 13.67

Changes in calibre

Table 13.2 **Causes of changes in calibre**

1. **Arterial attenuation**
   - Systemic hypertension (**Fig. 13.68**)
   - Retinal artery occlusion (**Fig. 13.69**)
   - Severe diffuse retinal disease such as retinitis pigmentosa (**Fig. 13.70**)
   - Cancer-associated retinopathy
2. **Combined venous and arterial dilatation and tortuosity**
   - Idiopathic (**Fig. 13.71**)
   - Optic nerve hypoplasia (**Fig. 13.72**)
   - Optic disc drusen (**Fig. 13.73**)
   - Retinopathy of prematurity (plus disease – **Fig. 13.74**)
   - Retinal capillary haemangioma (see Fig. 13.65)
   - Retinal racemose angioma (see Fig. 13.66)
   - Nanophthalmos
   - Maroteaux–Lamy syndrome
3. **Venous dilatation and/or tortuosity**
   - Inherited (**Fig. 13.75**)
   - Retinal vein occlusion (**Fig. 13.76**)
   - Preproliferative diabetic retinopathy (**Fig. 13.77**)
   - Hyperviscosity (**Fig. 13.78**)
   - Ocular ischaemic syndrome
   - Carotid–cavernous fistula
   - Primary antiphospholipid syndrome
   - Increased orbital pressure
   - Fabry disease
   - Mannosidosis

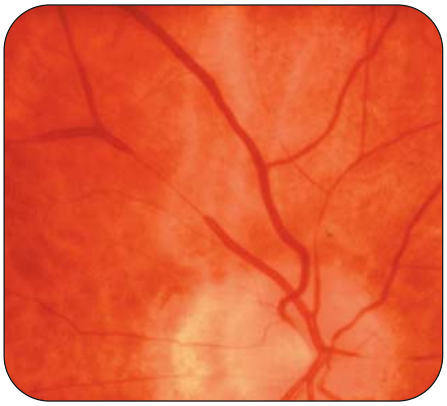
Fig. 13.68

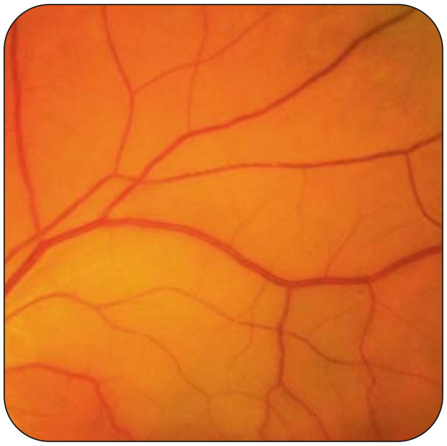
Fig. 13.69

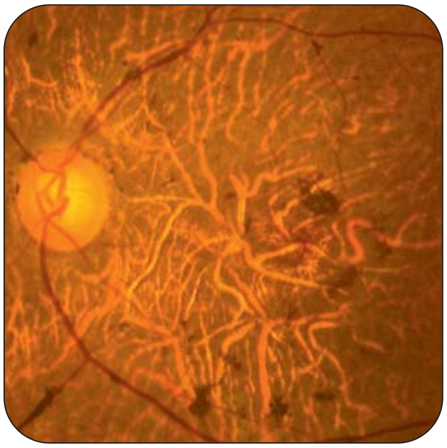
Fig. 13.70

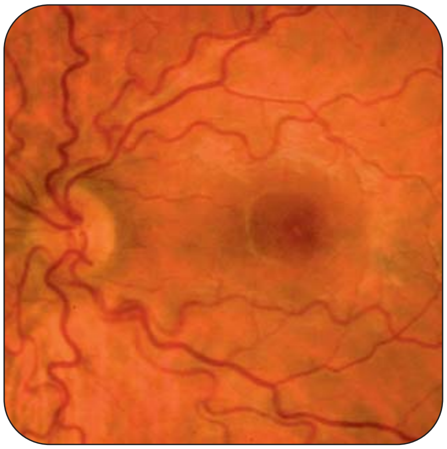
Fig. 13.71

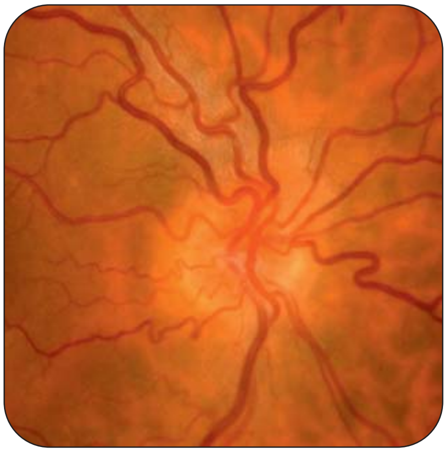
Fig. 13.72

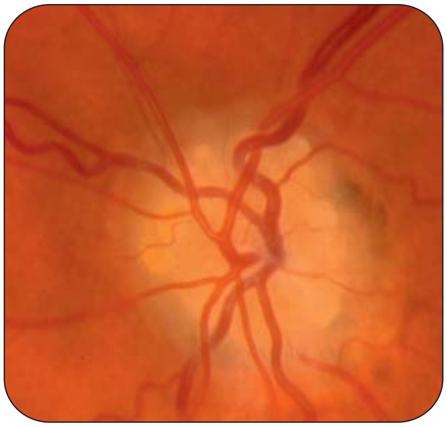
Fig. 13.73

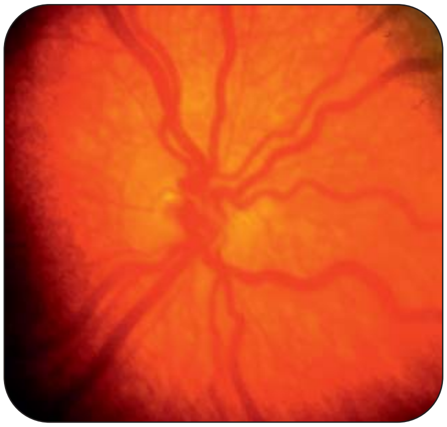
Fig. 13.74

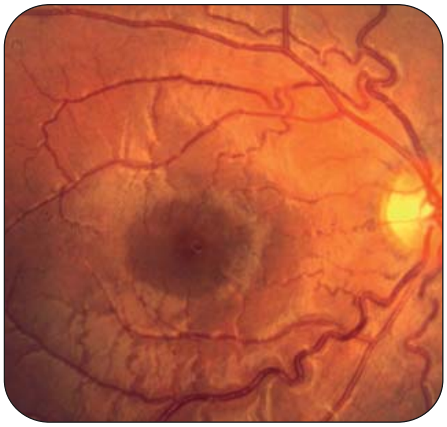
Fig. 13.75

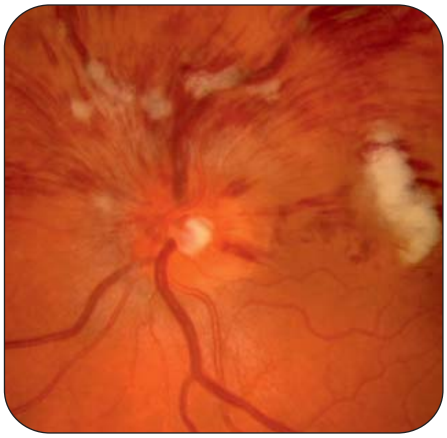
Fig. 13.76

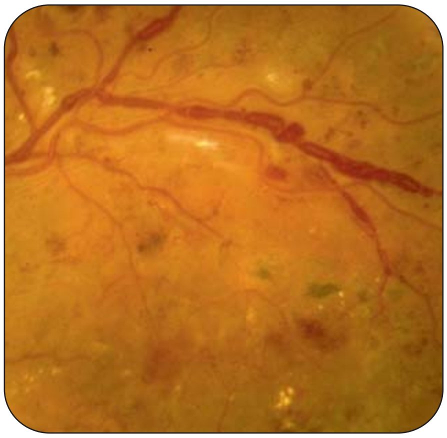
Fig. 13.77

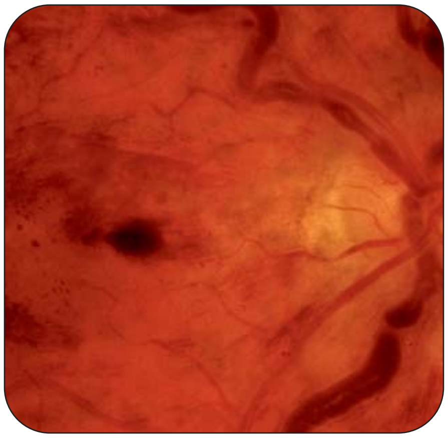
Fig. 13.78

INFLAMMATORY LESIONS

Vasculitis

Periphlebitis

Signs

- Patchy, fluffy, white haziness surrounding the venous blood column (**Fig. 13.79**) that may be associated with haemorrhage.
- Later there is venous sheathing (**Fig. 13.80**).
- FA shows vascular leakage and staining of the vessel wall (**Fig. 13.81**).

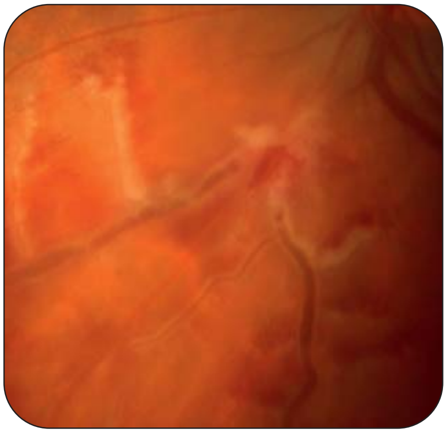

Fig. 13.79

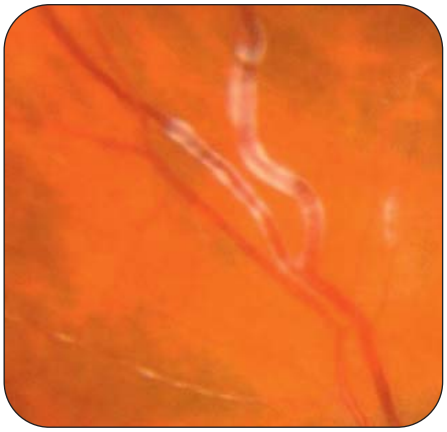

Fig. 13.80

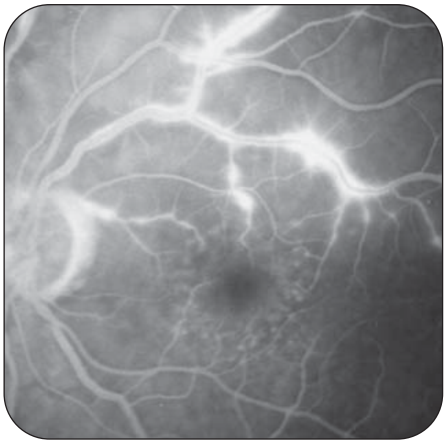

Fig. 13.81

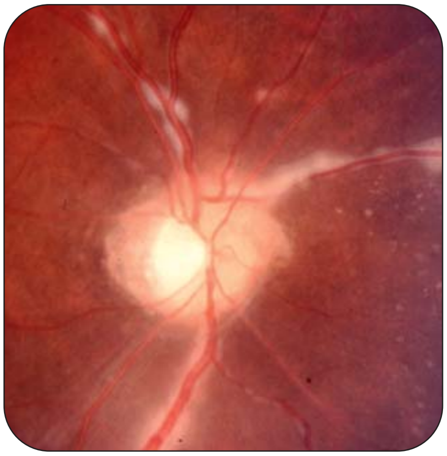

Fig. 13.82

Systemic associations

- Sarcoidosis – severe periphlebitis results in perivascular ‘candle-wax drippings’ (**Fig. 13.82**).
- Behçet disease – periphlebitis may be associated with haemorrhage and venous occlusion (**Fig. 13.83**); in some cases periarteritis may coexist (see below).

- AIDS – periphlebitis is frequently associated with CMV retinitis (**Fig. 13.84**).
- Tuberculosis – periphlebitis may lead to severe retinal ischaemia and secondary neovascularisation (**Fig. 13.85**).
- Multiple sclerosis – periphlebitis may be associated with intermediate uveitis.
- Cat-scratch fever.
- Whipple disease.
- Crohn disease.

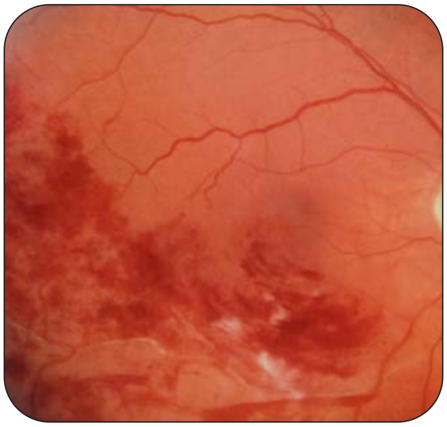

Fig. 13.83

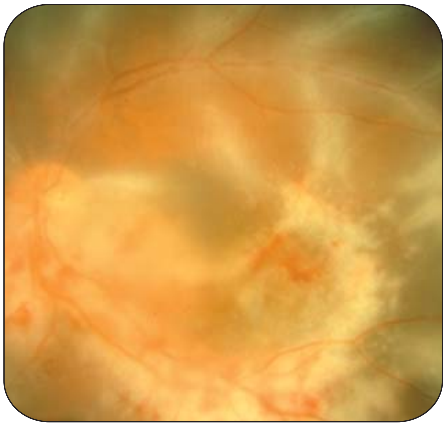

Fig. 13.84

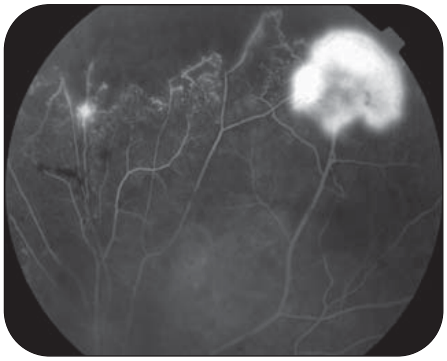

Fig. 13.85

Periarteritis

Signs

- Initially, patchy, fluffy white haziness surrounding the arteriolar blood column.
- Later there is arteriolar sheathing.
- Severe involvement may result in multiple branch artery occlusions (**Fig. 13.86**).
- FA shows staining of arterioles and variable occlusion.

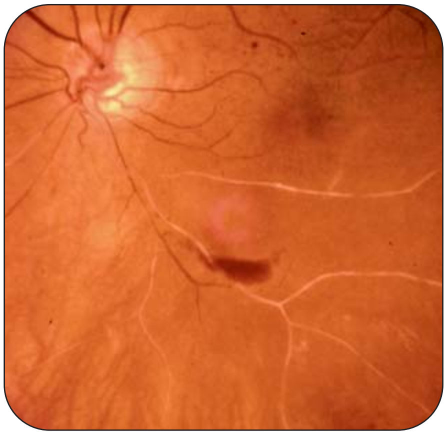

Fig. 13.86

Systemic associations

- Systemic lupus erythematosus.
- Polyarteritis nodosa.
- Dermatomyositis.
- Behçet disease (also periphlebitis).

- Acquired syphilis.
- Wegener granulomatosis.
- Kawasaki disease.
- Churg–Strauss syndrome.

Frosted branch angiitis

Definition

- A usually bilateral condition that may rarely be idiopathic and more commonly is secondary to CMV retinitis.

Signs

- Florid translucent perivascular sheathing of arteries and veins.
- Anterior uveitis, vitritis and retinal oedema are common.

Complications

- Venous occlusion (**Fig. 13.87**).
- Papillitis.
- Hard exudate formation.
- Retinal haemorrhage.

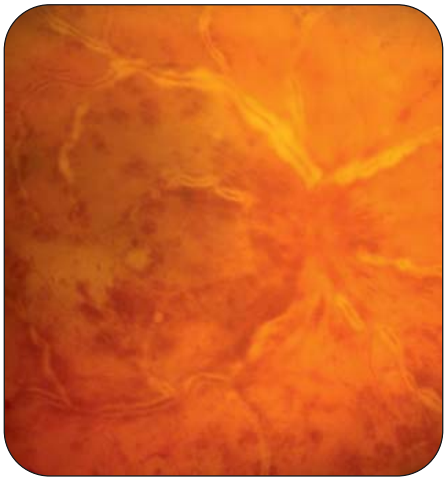

Fig. 13.87

Chorioretinitis with systemic associations

Toxoplasmosis

Definition

- A protozoan infection which typically affects one eye of a young individual.

Signs

- Anterior uveitis and vitritis.
- Solitary focal retinitis adjacent to an old scar (**Fig. 13.88**).
- Inactive lesions are atrophic scars with pigmentation (**Fig. 13.89**).

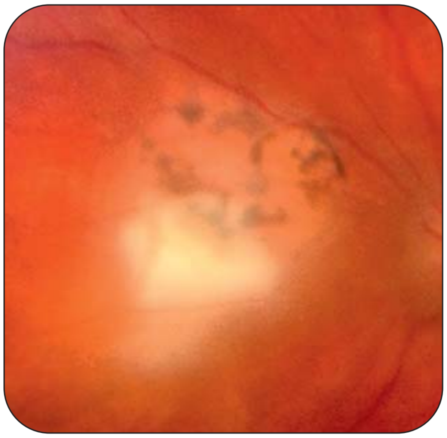

Fig. 13.88

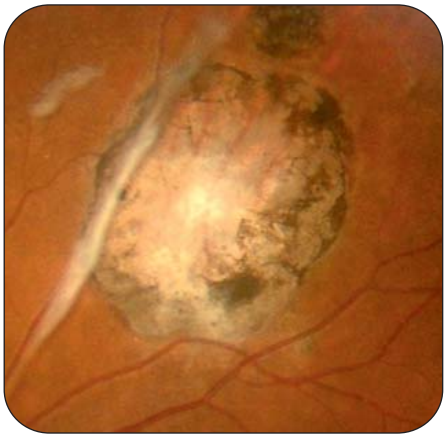

Fig. 13.89

Toxocariasis

Definition

- A very rare worm infection that is acquired in early childhood; ocular involvement is always unilateral.

Signs

- Posterior (**Fig. 13.90**) or peripheral granuloma that may be associated with 'dragging' of the disc (see Fig. 12.58).
- Chronic endophthalmitis (sees Figs 9.32 and 9.33).

Differential diagnosis

- Retinoblastoma may resemble a posterior-pole or peripheral granuloma.

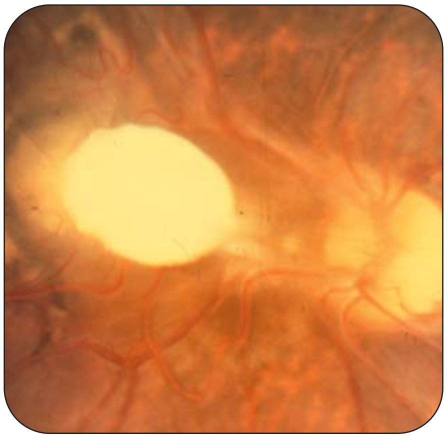

Fig. 13.90

Candidiasis

Definition

- A fungal infection which is associated with intravenous drug abuse, parenteral nutrition, chronic lung disease and neutropenia.

Signs

- Solitary or multiple retinal infiltrate.
- Vitritis and fluffy white 'cotton' balls (**Fig. 13.91**).

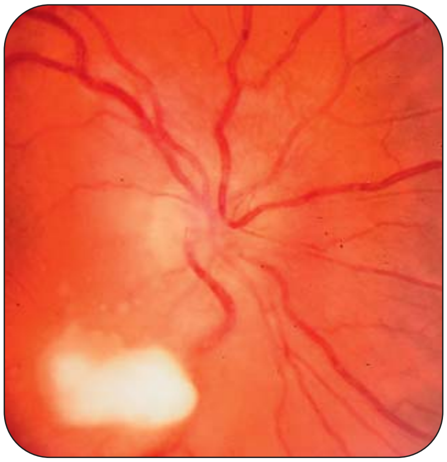

Fig. 13.91

Other causes of vitreous cotton balls

- Intermediate uveitis.
- Sarcoidosis.
- Lyme disease.
- Whipple disease.
- Seeding of endophytic retinoblastoma.

Sarcoidosis

Definition

- A common granulomatous multisystem disorder which affects the eyes in about 30% of cases.

Signs

- Anterior uveitis, which is most frequently chronic and granulomatous.
- Periphlebitis (see Fig. 13.82).
- Discrete, grey-white to waxy-yellow retinal and preretinal nodules (Lander sign – **Fig. 13.92**).

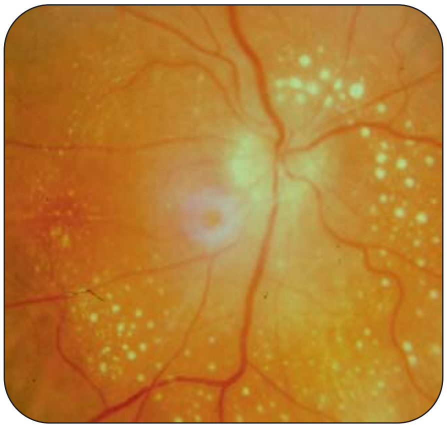

Fig. 13.92

- Multifocal choroidal infiltrates, which are more frequent in the inferior fundus (**Fig. 13.93**).
- Vitreous cotton balls.

Look for

- Black patient with facial palsy due to neurosarcoid (**Fig. 13.94**).

Differential diagnosis

- Multifocal choroiditis with panuveitis.
- Birdshot retinochoroidopathy.

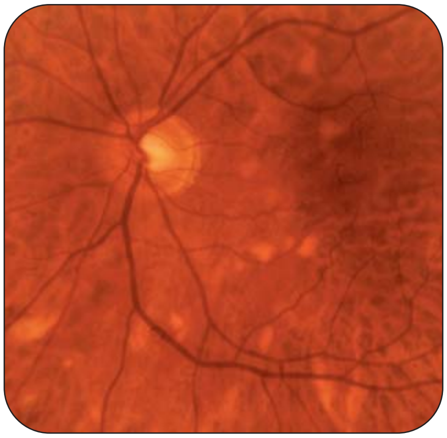
Fig. 13.93

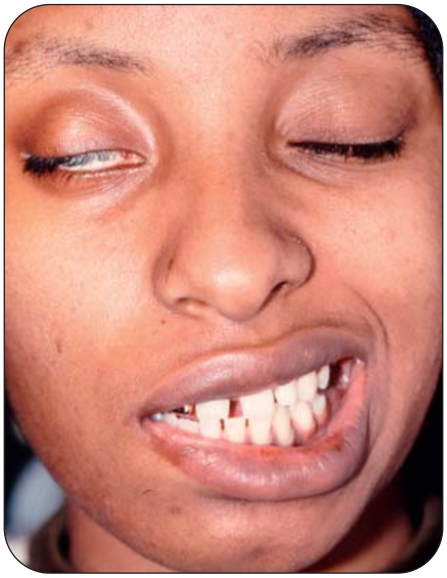
Fig. 13.94

Behçet disease

Definition

- An uncommon multisystem disorder which is frequently associated with ocular manifestations which are usually bilateral.

Signs

- Acute anterior uveitis with hypopyon but a relatively white eye (see Fig. 8.17).
- Vitritis.
- White, necrotic, superficial retinal infiltrates (**Fig. 13.95**).

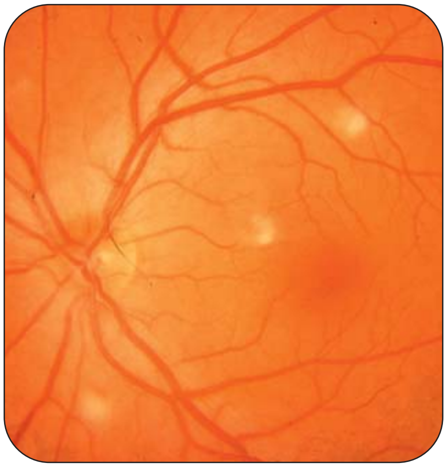
Fig. 13.95

Complications

- Vasculitis and venous occlusion (see Fig. 13.83).
- Macular oedema.
- Vascular attenuation and optic atrophy in end-stage disease (**Fig. 13.96**).

Look for

- Superficial thrombophlebitis (**Fig. 13.97**).

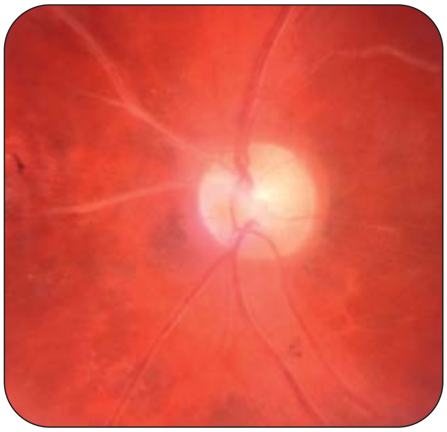

Fig. 13.96

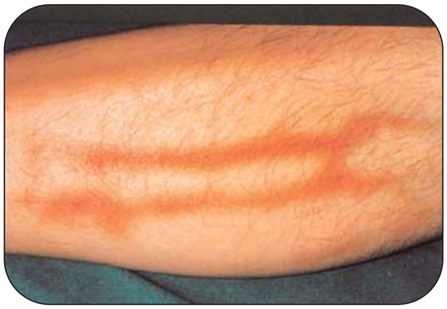

Fig. 13.97

Vogt–Koyanagi–Harada syndrome (VKH)

Definition

- An uncommon idiopathic condition which typically affects pigmented individuals; bilateral ocular involvement is the rule.

Signs

- Chronic granulomatous anterior uveitis.
- Multifocal choroiditis followed by detachments of the sensory retina (**Fig. 13.98A**).
- FA venous phase shows multiple hyperfluorescent spots (**Fig. 13.98B**); late phase manifests pooling of dye under the serous retinal detachments (**Fig. 13.98C**).

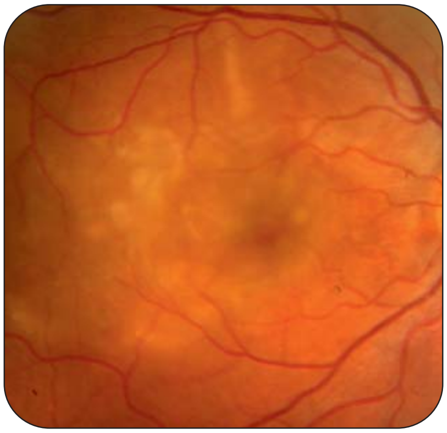

Fig. 13.98A

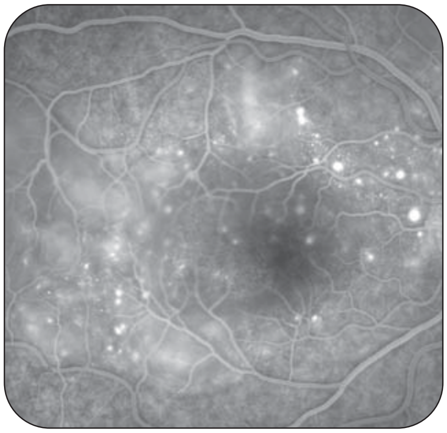

Fig. 13.98B

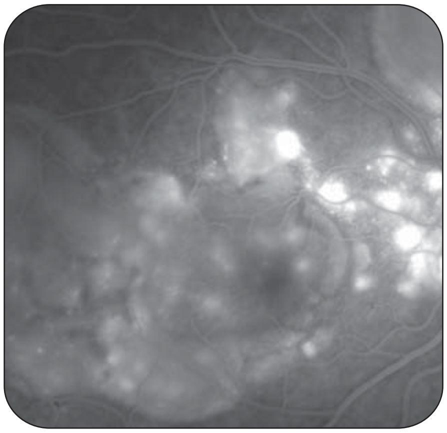

Fig. 13.98C

- Exudative retinal detachment.
- Yellow-white, well-circumscribed atrophic spots, giving rise to a ‘sunset glow’ appearance in inactive disease (**Fig. 13.99**).

Look for

- Vitiligo (**Fig. 13.100**).

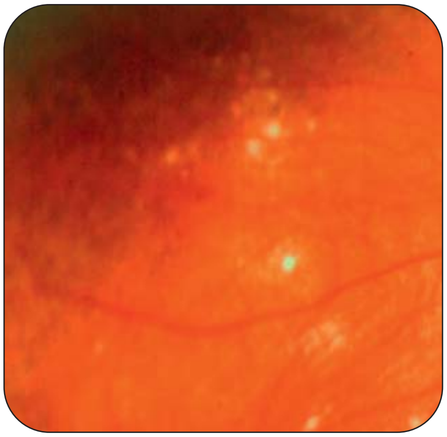

Fig. 13.99

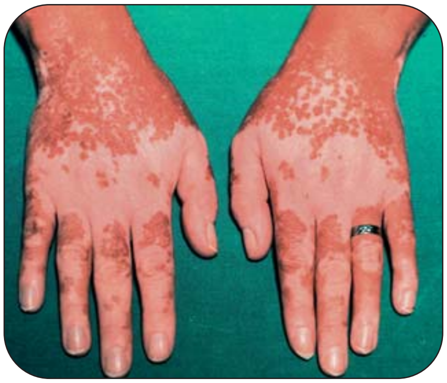

Fig. 13.100

Tuberculous

Signs

- Chronic granulomatous anterior uveitis.
- Solitary (**Fig. 13.101**) or less commonly multifocal choroiditis.
- Retinal vasculitis (see Fig. 13.85).

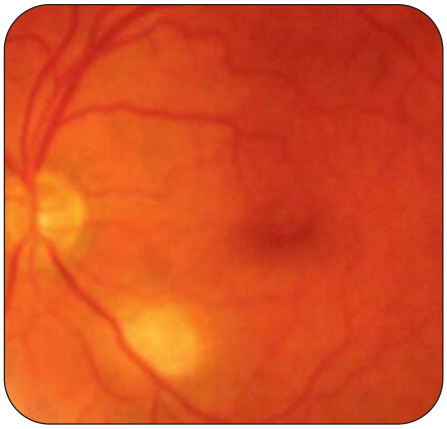

Fig. 13.101

Acquired syphilis

Signs

- Anterior uveitis – acute or granulomatous.
- Neuroretinitis (see Fig. 13.35).
- Solitary or multifocal, large, deep, placoid lesions with faded centres, at the posterior pole (**Fig. 13.102**).
- Hypopigmented chorioretinal scars and pigment migration in inactive disease (**Fig. 13.103**).

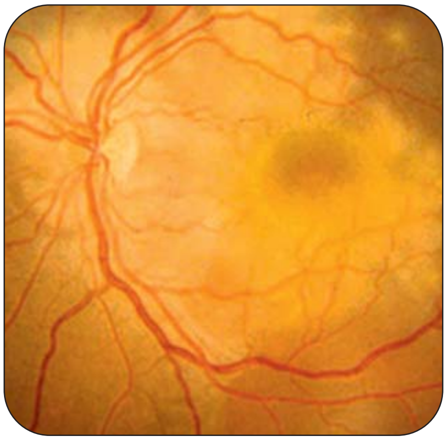

Fig. 13.102

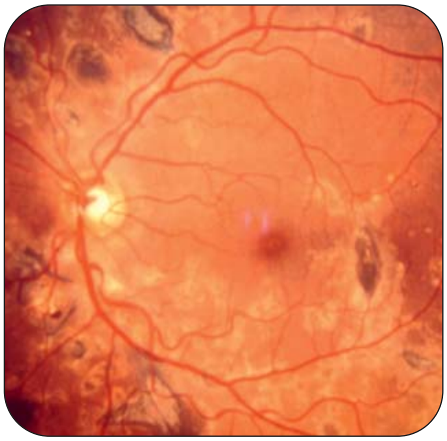

Fig. 13.103

Choroidal pneumocystosis

Definition

- A rare infection with *Pneumocystis jiroveci* which is a nonseptate mould that affects patients with AIDS.

Signs

- Bilateral, large, multifocal, slightly elevated, round or oval, plaque-like, yellowish choroidal lesions predominantly involving the posterior pole (**Fig. 13.104**).
- Vitritis is absent.

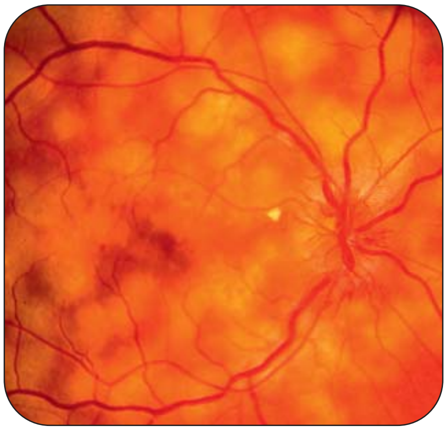

Fig. 13.104

Chorioretinitis without systemic associations

Sympathetic ophthalmitis

Definition

- A bilateral granulomatous panuveitis which occurs following penetrating trauma.

Signs

- Chronic granulomatous anterior uveitis.
- Extensive multifocal choroiditis (**Fig. 13.105**).

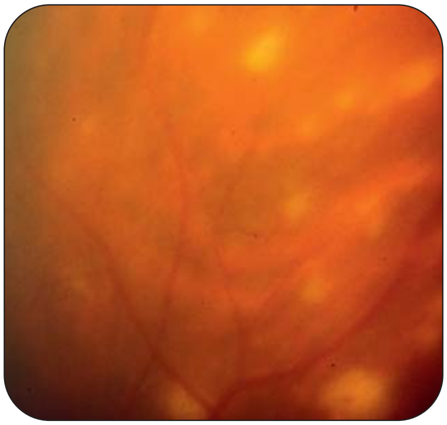

Fig. 13.105

Multifocal choroiditis with panuveitis syndrome

Definition

- A unilateral or bilateral, frequently recurrent condition, that typically affects middle-aged women.

Signs

- Anterior uveitis and vitritis.
- Small, discrete, old and fresh choroidal lesions located at the posterior pole and/or the midperiphery (**Fig. 13.106**).

Complications

- CNV is uncommon.
- Subretinal fibrosis is rare.

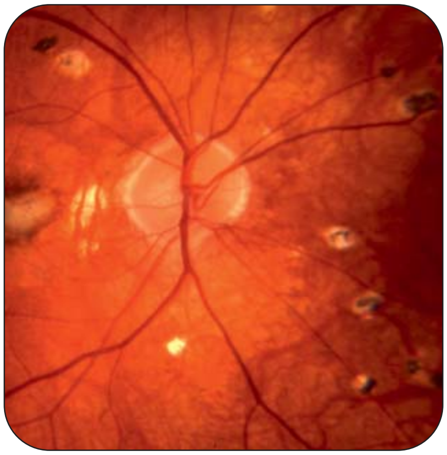

Fig. 13.106

Differential diagnosis

- Sarcoidosis.
- Acute lesions may resemble punctate inner choroidopathy but the latter is not associated with vitritis.
- POHS may show similar lesions but is not associated with vitritis (see next).

Presumed ocular histoplasmosis syndrome

Definition

- A fungal infection which is common in certain parts of the USA but rare in Europe.

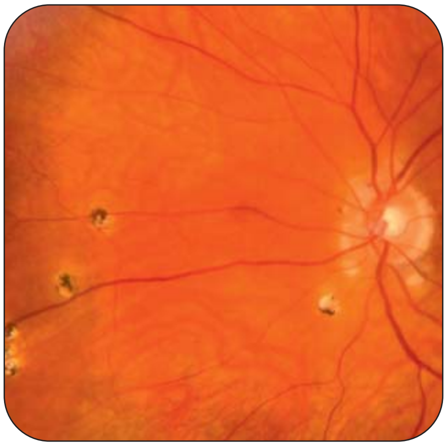
Fig. 13.107

Signs

- No vitritis.
- Peripapillary atrophy and small, discrete, yellowish lesions often associated with pigment scattered in the posterior fundus and mid-periphery (**Fig. 13.107**).

Complications

- CNV is common.

Differential diagnosis

- Paving-stone degeneration may resemble the peripheral atrophic lesions.
- Degenerative myopia may cause both the atrophic lesions and CNV.
- Punctate inner choroidopathy (see below).
- Inactive multifocal choroiditis with panuveitis syndrome.

Punctate inner choroidopathy

Definition

- A usually eventually bilateral chronic condition that typically affects young myopic women.

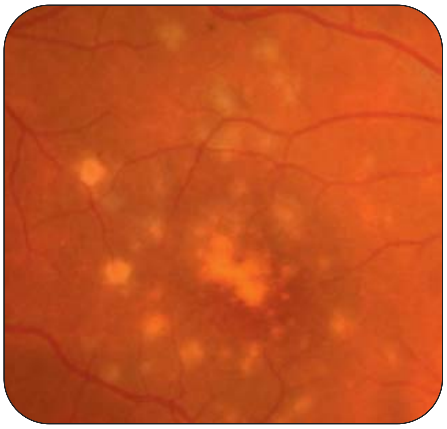
Fig. 13.108

Signs

- No vitritis.
- Small, yellow, indistinct choroidal lesions all of same age at the posterior pole (**Fig. 13.108**).

Complications

- CNV is common.

Birdshot retinochoroidopathy

Definition

- A usually bilateral chronic condition that typically affects middle-aged individuals who carry HLA-A29.

Signs

- Moderate vitritis.
- Moderate size, creamy-yellow, deep ovoid spots with indistinct edges, which radiate from the optic disc towards the equator (**Fig. 13.109**).
- ICG shows well-defined hypofluorescent spots in the early phase (**Fig. 13.110**) that later become hyperfluorescent.

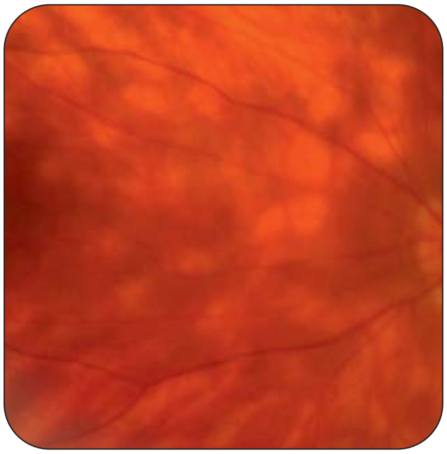
Fig. 13.109

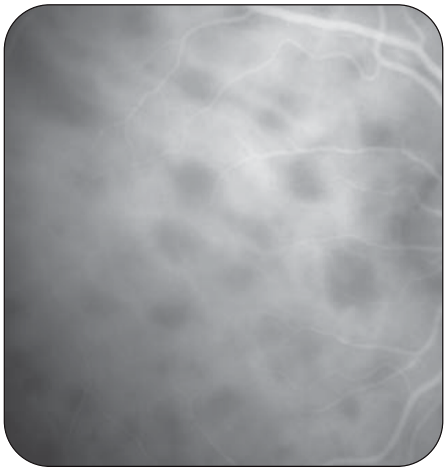
Fig. 13.110

Acute posterior multifocal placoid pigment epitheliopathy

Definition

- A bilateral, self-limiting, condition which typically affects young adults.

Signs

- Mild vitritis.
- Large, cream-coloured, placoid lesions at the level of the RPE at the posterior pole and midperiphery (**Fig. 13.111A**).
- FA initially shows dense hypofluorescence (**Fig. 13.111B**) followed by diffuse hyperfluorescence (**Fig. 13.111C**).

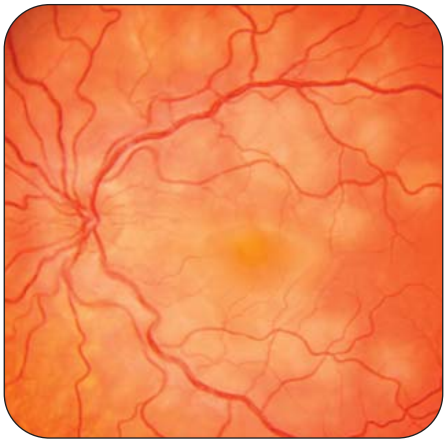
Fig. 13.111A

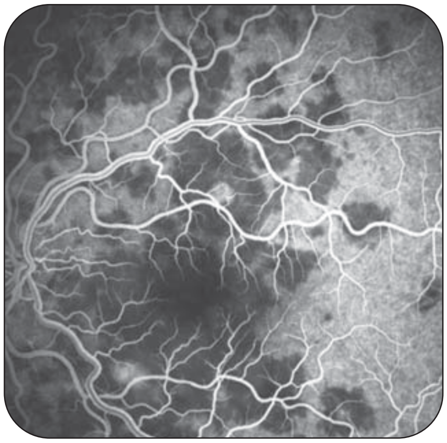
Fig. 13.111B

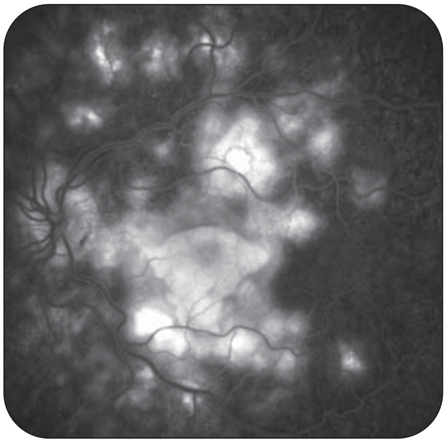
Fig. 13.111C

Serpiginous choroidopathy

Definition

- A bilateral chronic condition which typically affects middle-aged individuals.

Signs

- Mild vitritis.
- Multiple choroidal lesions with hazy borders that typically start near the disc and the spread outwards (**Fig. 13.112**).

Complications

- CNV is uncommon.

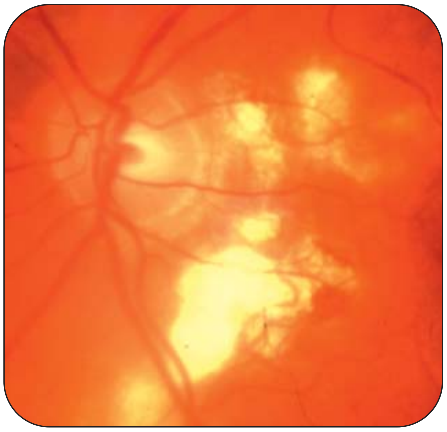

Fig. 13.112

Necrotising retinitis

Cytomegalovirus retinitis

Definition

- A chronic, often bilateral, viral infection that affects patients with AIDS.

Signs

- Mild vitritis.
- Yellow-white areas of retinitis and haemorrhage which typically start at the posterior pole and then frequently spread along the vascular arcades (**Fig. 13.113**).

Complications

- Retinal atrophy.
- Retinal detachment.

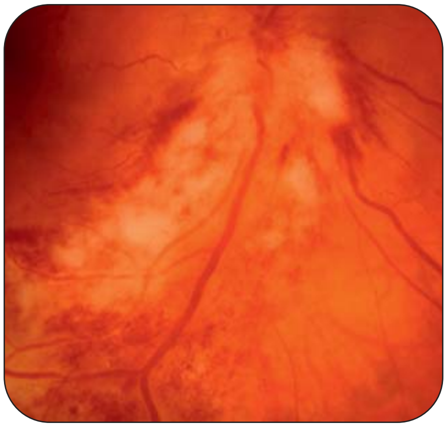

Fig. 13.113

Acute retinal necrosis

Definition

- An infection caused by the varicella-zoster or herpes simplex virus which is often bilateral and usually affects otherwise healthy individuals.

Signs

- Anterior uveitis and vitritis.
- Confluent, peripheral occlusive necrotising vasculitis.
- Progression is faster than in cytomegalovirus retinitis but the macula is usually spared till late (**Fig. 13.114**).

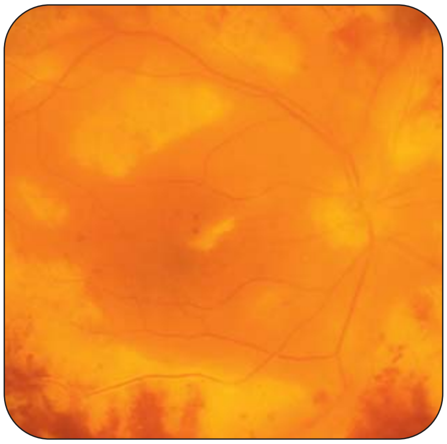

Fig. 13.114

Differential diagnosis

- Intraocular lymphoma may be associated with vitreous cells and diffuse retinal involvement.

Progressive outer retinal necrosis

Definition

- An infection caused by varicella-zoster virus which is often bilateral and predominantly affects patients with AIDS.

Signs

- No vitritis.
- Necrotising retinitis that starts in the periphery or the posterior pole.
- Rapidly progresses with early macular involvement (**Fig. 13.115**).

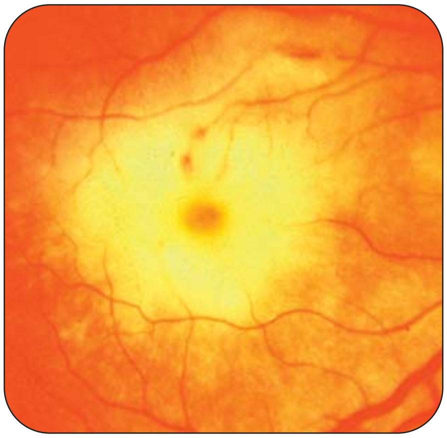

Fig. 13.115

MACULAR LESIONS

Oedema

Macular oedema is characterised by increased fluid within the sensory retina that may assume a cystoid or non-cystoid configuration.

Non-cystoid

Signs

- Retinal thickening at the posterior pole.
- FA shows intraretinal leakage during the late phase (**Fig. 13.116**).
- OCT shows macular thickening with loss of the foveal pit (**Fig. 13.117**).

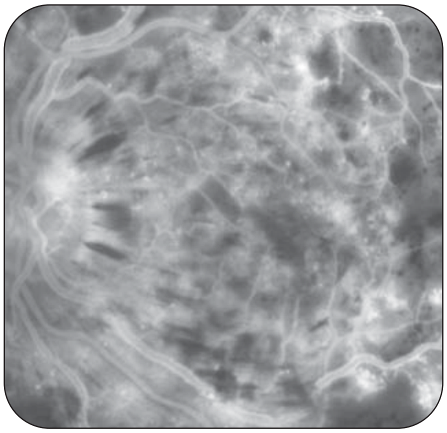

Fig. 13.116

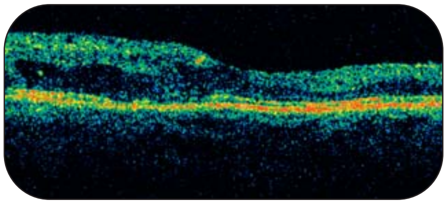

Fig. 13.117

Cystoid

Signs

- Thickening of the fovea associated with microcysts (**Fig. 13.118**).
- Best seen with red-free light (**Fig. 13.119**).
- FA shows a flower-petal pattern of leakage at the fovea during the late phases (**Fig. 13.120**).
- OCT shows cystoid spaces within thickened retina (**Fig. 13.121**).

Differential diagnosis

- Foveal schisis which is a bilateral XL hereditary condition characterised by a 'bicycle-wheel' appearance (**Fig. 13.122**); 50% of cases have associated peripheral retinoschisis (**Fig. 13.123**).

Table 13.3 **Causes of macular oedema**

1. **Retinal vascular disease**
   - Background diabetic retinopathy
   - Retinal vein occlusion
2. **Iatrogenic**
   - Cataract surgery (Irvine–Gass syndrome)
   - YAG laser capsulotomy
   - Retinal laser photocoagulation or cryotherapy
   - Topical 2% epinephrine in aphakia
   - Topical latanoprost – very rare
3. **Chronic intraocular inflammation**
   - Anterior uveitis
   - Intermediate uveitis
   - Birdshot retinochoroidopathy
   - Panuveitis
   - Infectious endophthalmitis
4. **Secondary to other maculopathies**
   - CNV
   - Preretinal macular fibrosis
   - Vitreomacular traction syndrome
5. **Associated with hereditary dystrophies**
   - Retinitis pigmentosa
   - Gyrate atrophy
6. **Miscellaneous**
   - High doses systemic of nicotinic acid
   - Familial dominant

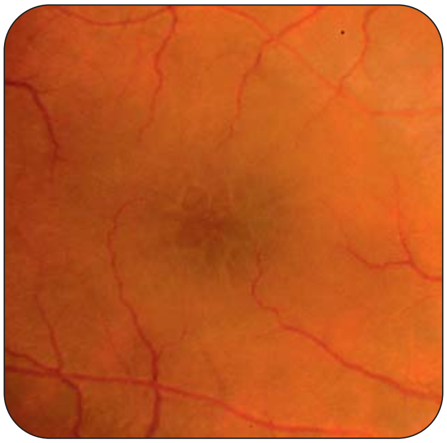
Fig. 13.118

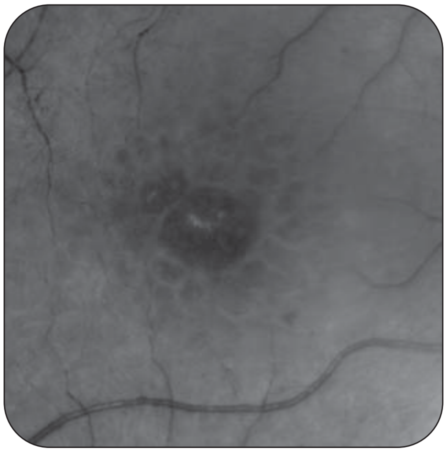
Fig. 13.119

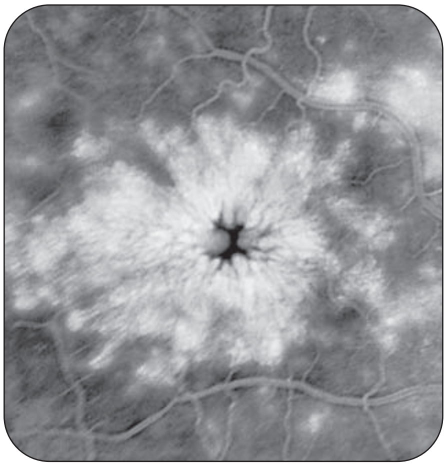
Fig. 13.120

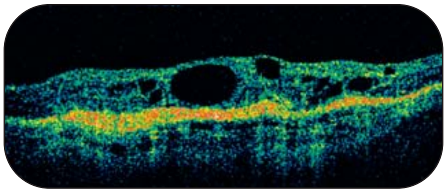
Fig. 13.121

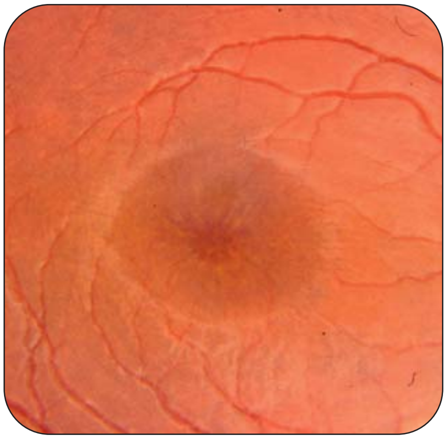
Fig. 13.122

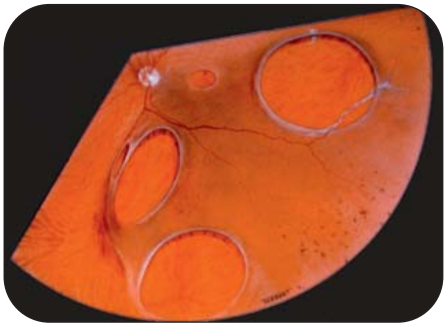
Fig. 13.123

Serous detachment

In serous detachment there is a separation of the sensory retina from the RPE by fluid; the causes are the following:

Idiopathic central serous retinopathy

Signs

- Unilateral, shallow elevation of the sensory retina at the posterior pole, with indistinct borders (**Fig. 13.124**).
- Associated tiny subretinal precipitates may be seen.
- FA shows progressive 'smoke-stack' hyperfluorescence under the sensory retina (**Fig. 13.125**). An expanding 'ink-blot' leak is less common (**Fig. 13.126**).
- OCT shows shallow separation of the sensory retina from the RPE (**Fig. 13.127**).

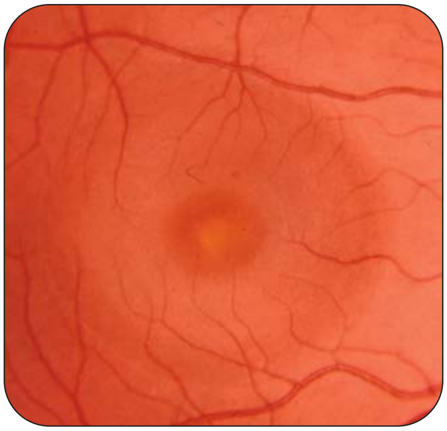

Fig. 13.124

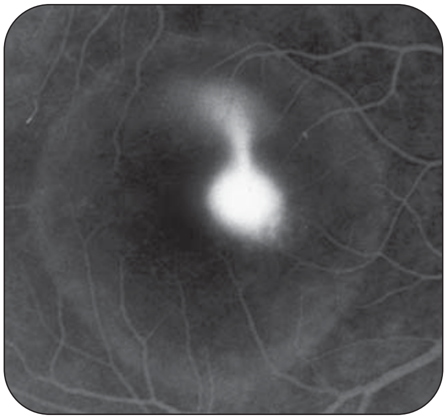

Fig. 13.125

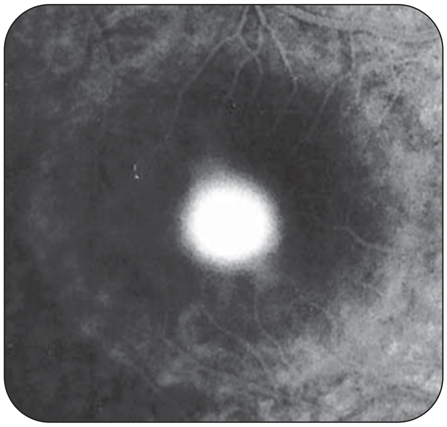

Fig. 13.126

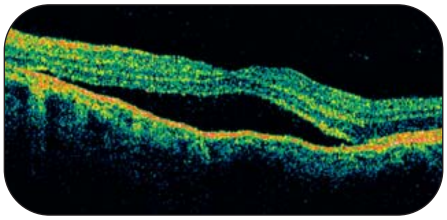

Fig. 13.127

Congenital optic disc pit

Signs

- Macular serous detachment develops in about 45% of eyes with disc pits.
- Initially there is a schisis-like separation of the inner retinal layers which communicates with the pit (**Fig. 13.128**).
- Separation of the outer layers develops later (**Fig. 13.129**).

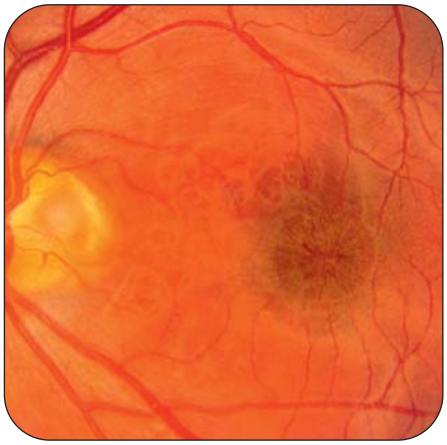

Fig. 13.128

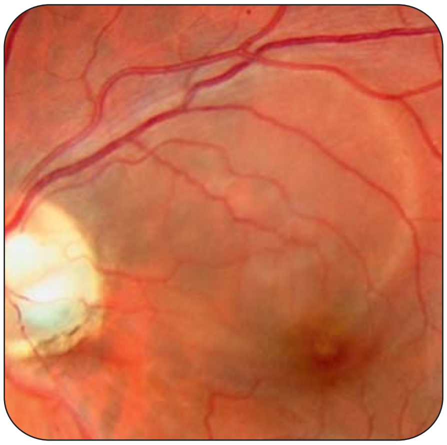

Fig. 13.129

Other causes

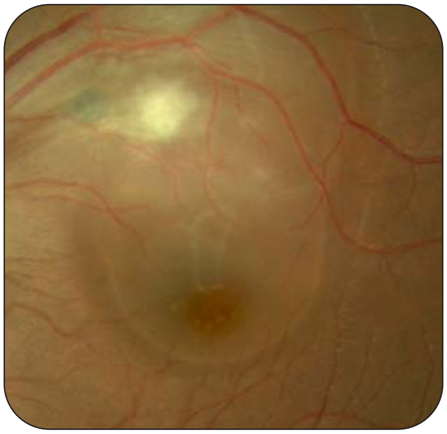

Fig. 13.130

- Retinitis (**Fig. 13.130**) may occasionally cause a serous detachment.
- Harada disease (see Fig. 13.98).

Detachment of the RPE

Detachment of the RPE typically affects elderly patients with age-related macular degeneration.

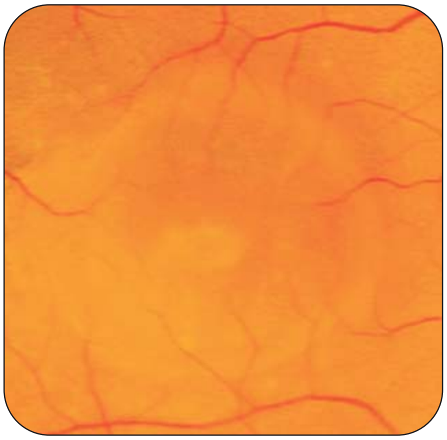

Fig. 13.131

Signs

- Unilateral, sharply circumscribed, dome-shaped elevation at the posterior pole of varying size (**Fig. 13.131**).
- Sub-RPE fluid may be clear or cloudy.
- FA shows an area of early hyperfluorescence which increases in intensity but not in size during the later phases (**Fig. 13.132**).
- ICG shows hypofluorescence with a faint hyperfluorescent outline (**Fig. 13.133**).
- OCT shows a dome-shaped separation of the RPE from Bruch membrane (**Fig. 13.134**).

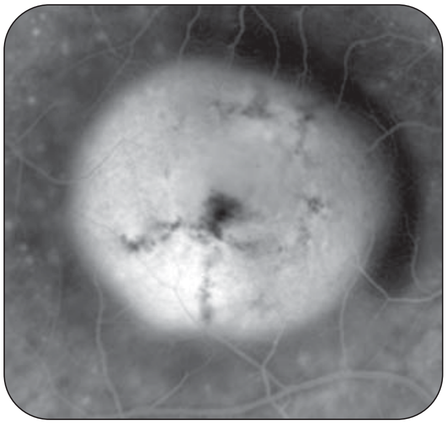

Fig. 13.132

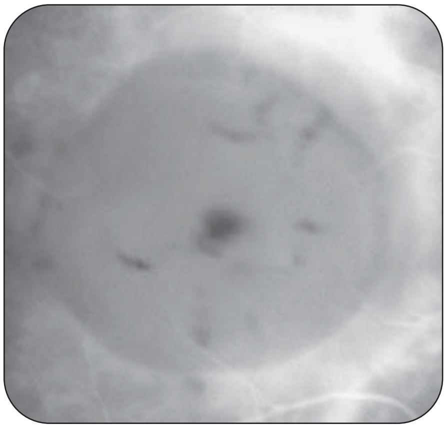

Fig. 13.133

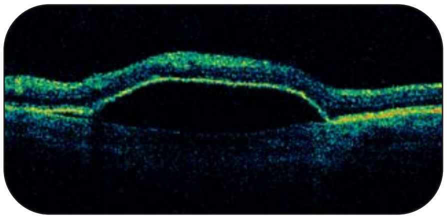

Fig. 13.134

Choroidal neovascularisation

In choroidal neovascularisation (CNV) new vessels originating from the choriocapillaris grow through defects in Bruch membrane under the RPE (type 1 CNV) or into the subretinal space (type 2), or both.

Signs

In chronological order:

- Leaking from CNV causing macular oedema, hard exudates and haemorrhages (**Fig. 13.135**).
- Subretinal and sub-RPE bleeding (**Fig. 13.136**).
- Subretinal (disciform) scarring (**Fig. 13.137**).

FA

- 'Lacy' pattern of hyperfluorescence in the very early phase (**Fig. 13.138A**).
- Bright hyperfluorescence during peak dye transit (**Fig. 13.138B**).
- Late leakage into the subretinal space and around the CNV (**Fig. 13.138C**).

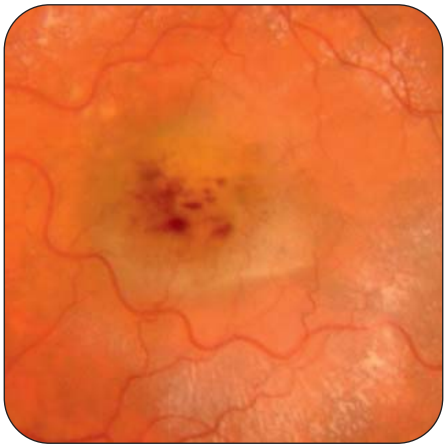

Fig. 13.135

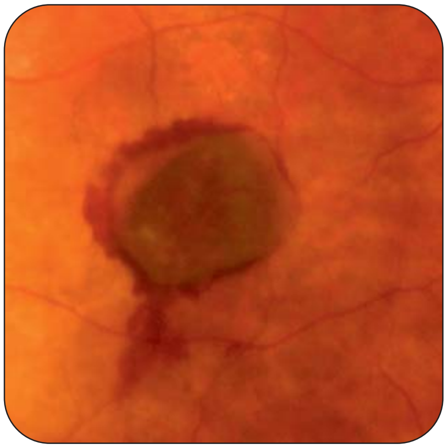

Fig. 13.136

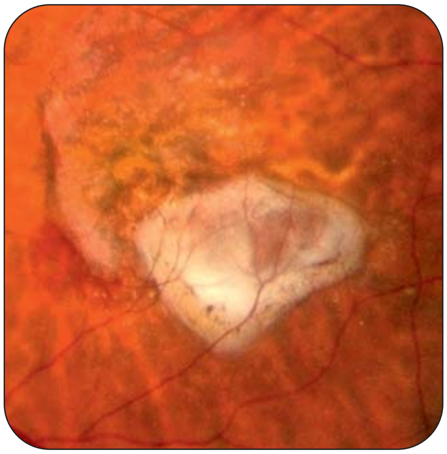

Fig. 13.137

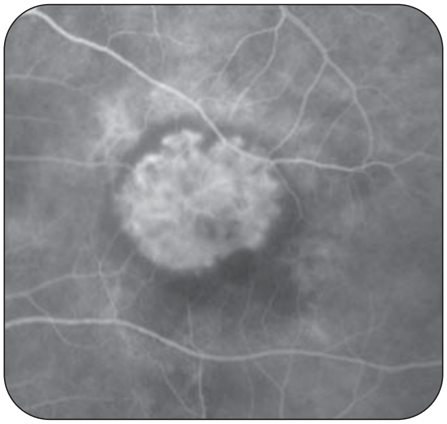

Fig. 13.138A

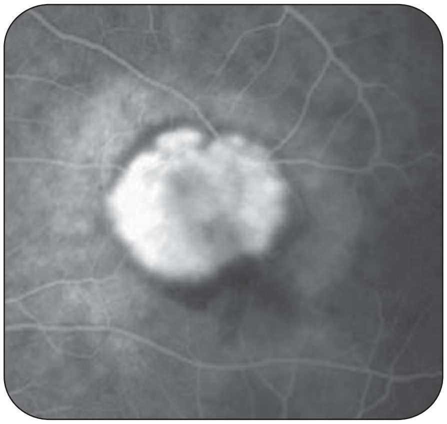

Fig. 13.138B

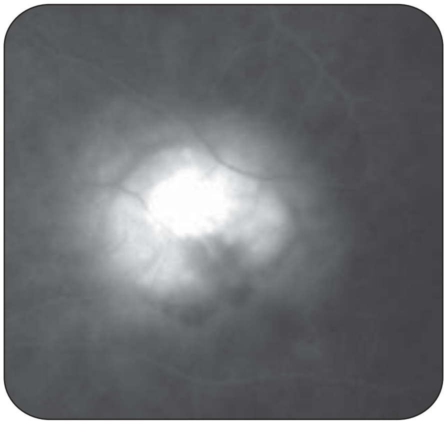

Fig. 13.138C

Degenerative causes

- Soft drusen (**Fig. 13.139**). FA shows corresponding hyperfluorescence due to RPE window defects (**Fig. 13.140**).
- Lacquer cracks in high myopia (**Fig. 13.141**).
- Angioid streaks are linear, grey or dark-red lesions with serrated edges that radiate outwards from the disc (**Fig. 13.142**). FA shows corresponding hyperfluorescence and is useful in detecting CNV which may be multifocal (**Fig. 13.143**). Pseudoxanthoma elasticum is present in about 50% of cases (**Fig. 13.144**); other conditions are less common (Table 13.4).
- Sorsby pseudo-inflammatory macular dystrophy.

Table 13.4 **Systemic associations of angioid streaks**

1. **Skin**
   - Pseudoxanthoma elasticum
   - Ehlers–Danlos syndrome
   - Senile elastosis
2. **Bone**
   - Paget disease
   - Acromegaly
   - Calcinosis
3. **Blood**
   - Sickle cell anaemia
   - Thalassaemia
   - Abetalipoproteinaemia
   - Hereditary spherocytosis

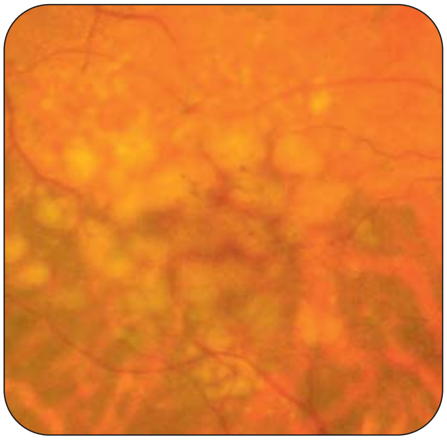

Fig. 13.139

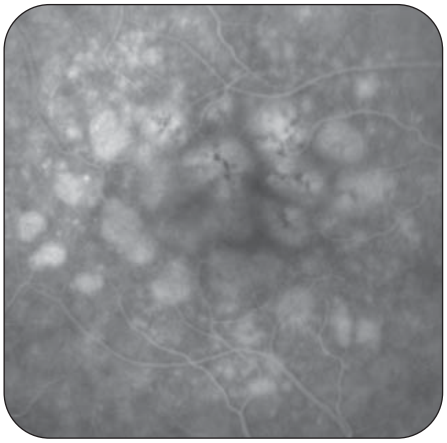

Fig. 13.140

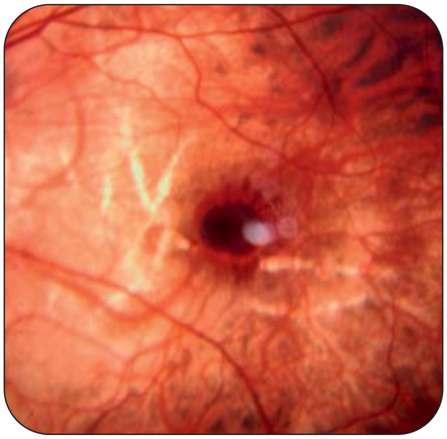

Fig. 13.141

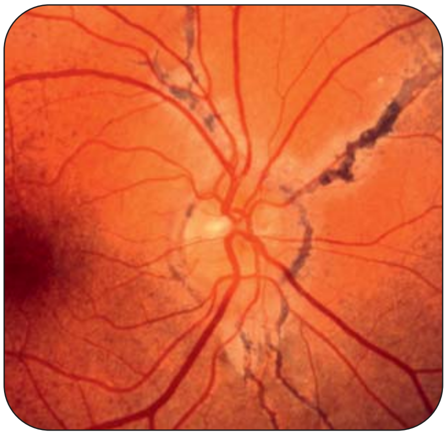

Fig. 13.142

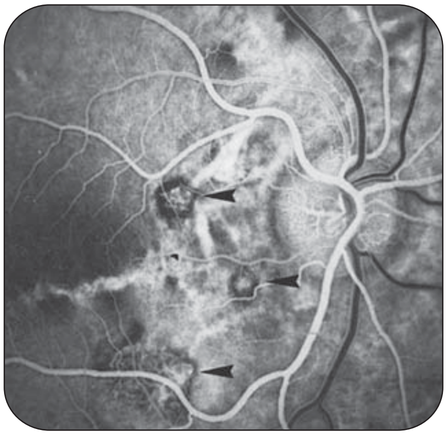

Fig. 13.143

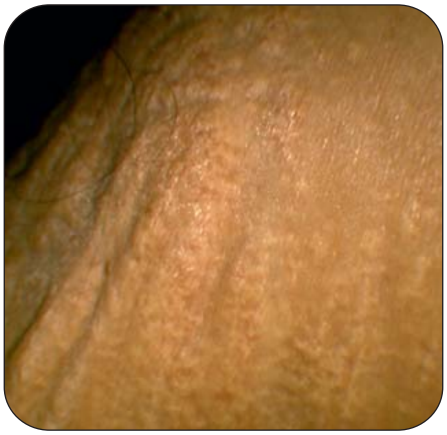

Fig. 13.144

Inflammatory causes

- Punctate inner choroidopathy (see Fig. 13.108) is frequently associated with CNV which may be multifocal (**Fig. 13.145**).
- Presumed ocular histoplasmosis syndrome (**Fig. 13.146**) is a common association.
- Rubella retinopathy.
- Serpiginous choroidopathy.
- Birdshot retinochoroidopathy.
- Multifocal choroiditis with panuveitis syndrome.

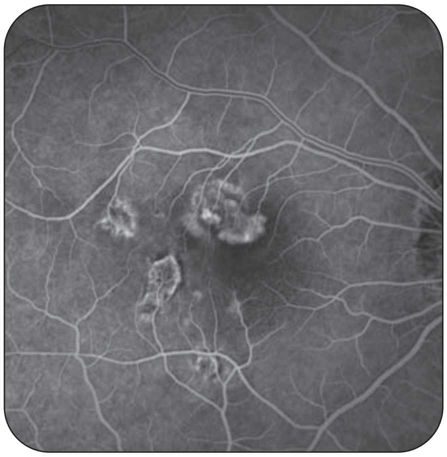

Fig. 13.145

Miscellaneous causes

- Choroidal naevus.
- Choroidal osteoma.
- Traumatic choroidal rupture is a rare cause (**Fig. 13.147**).
- Optic disc drusen is an occasional association of juxtapapillary CNV (**Fig. 13.148**).
- Best disease.

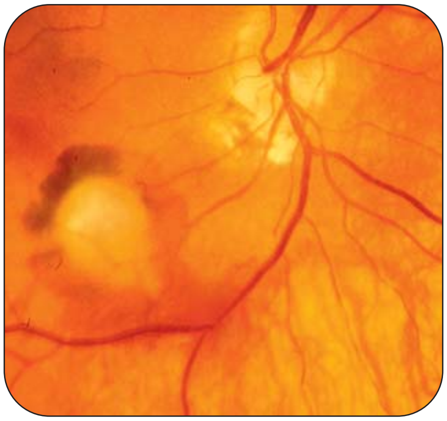

Fig. 13.146

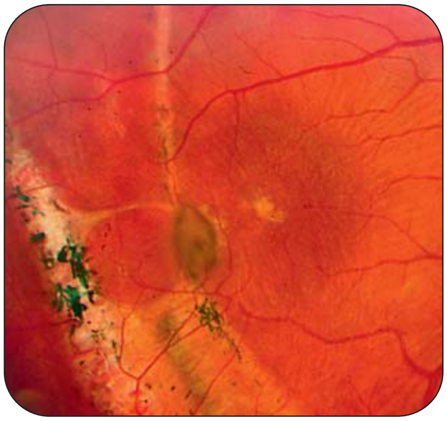

Fig. 13.147

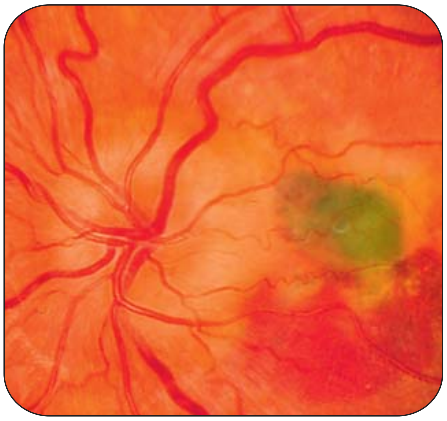

Fig. 13.148

Atrophic maculopathy

Dry age-related macular degeneration

Definition

- The most common form of age-related macular degeneration in which involvement is bilateral but frequently asymmetric.

Signs

- Sharply delineated areas of hypopigmentation or depigmentation in which choroidal vessels are more visible (**Fig. 13.149**).

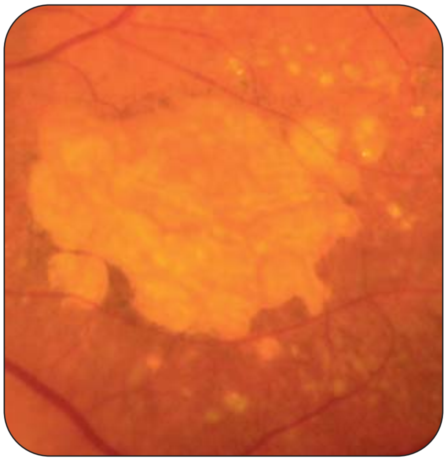

Fig. 13.149

Myopic maculopathy

Definition

- A usually bilateral but asymmetric condition which affects highly myopic adults.

Signs

- Roundish yellow-white areas at the macula due to focal chorioretinal atrophy that may be associated with a pigmented (Fuchs) spot (**Fig. 13.150**).
- Lacquer cracks at the posterior pole (see Fig. 13.141).
- Macular hole which may give rise to retinal detachment.
- Tilted disc and peripapillary atrophy.
- Peripheral chorioretinal atrophy, lattice degeneration and retinal breaks.
- Vitreous degeneration.

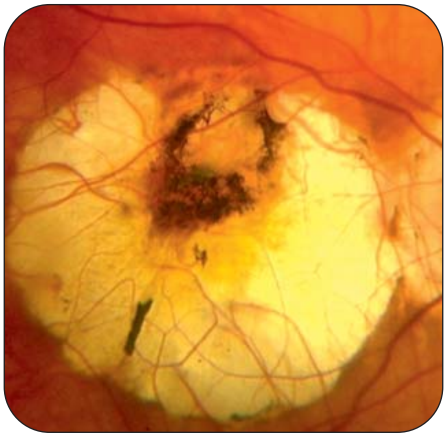

Fig. 13.150

Stargardt macular dystrophy

Definition

- An AR condition which starts during the teen years and results in early severe visual loss.

Signs

Three patterns may be seen:

- Isolated atrophic maculopathy (**Fig. 13.151**).
- Atrophic maculopathy surrounded by fish-tail-shaped flecks (**Fig. 13.152**).
- Atrophic maculopathy associated with diffuse flecks (**Fig. 13.153**).

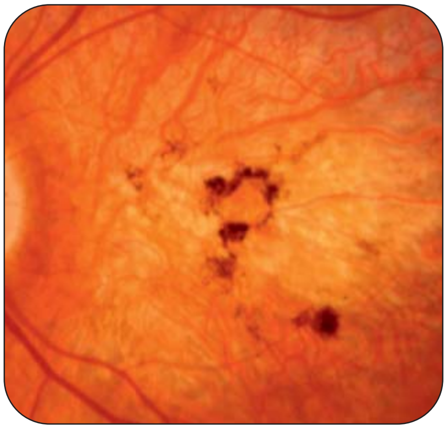

Fig. 13.151

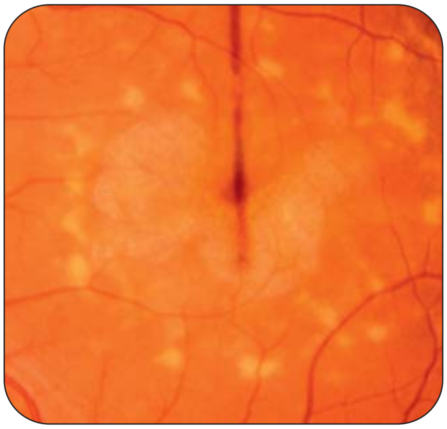

Fig. 13.152

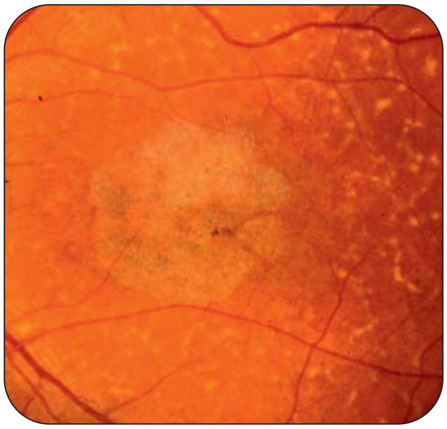

Fig. 13.153

Central areolar choroidal dystrophy

Definition

- An AD condition which causes loss of central vision during the 5th decade.

Signs

- Circumscribed, atrophic macular lesion between one and three disc diameters in size not associated with drusen (**Fig. 13.154**).

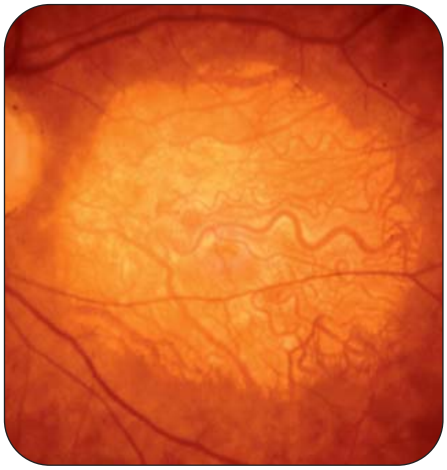

Fig. 13.154

Bull's eye maculopathy

Signs

- Central foveolar hyperpigmentation surrounded by an oval depigmented zone which is itself outlined by a ring of hyperpigmentation (**Fig. 13.155**).
- FA shows an RPE window defect corresponding to the depigmented zone (**Fig. 13.156**).

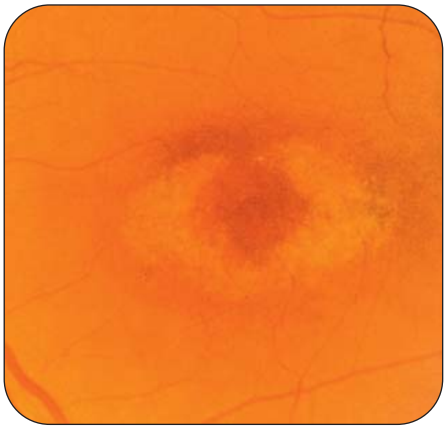

Fig. 13.155

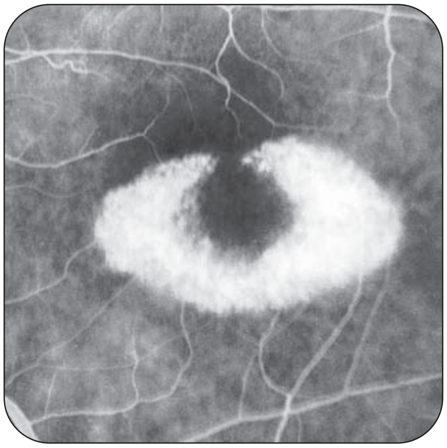

Fig. 13.156

Causes

a. *Chloroquine* may occasionally give rise to bull's eye maculopathy but hydroxychloroquine very rarely if ever causes retinal toxicity.

b. *Cone dystrophy* is a rare condition in which bull's eye maculopathy may be associated with temporal disc pallor, retinal vascular attenuation and mild peripheral pigmentary changes (**Fig. 13.157**).

c. *Benign concentric annular macular dystrophy* is an AD condition which may cause mild impairment of central vision.

d. *Inverse RP* in which the pigmentary changes are confined to the posterior pole.

e. *Fenestrated sheen macular dystrophy* is an AD condition in which bull's eye maculopathy is a late feature.

f. *Leber congenital amaurosis* is an AR retinal dystrophy that causes perinatal blindness in which bull's eye maculopathy is one of many fundus features. Oculodigital syndrome (**Fig. 13.158**), roving nystagmus and poor pupillary light reactions are characteristic.

g. *Stargardt disease* (see above) may occasionally assume a bull's eye configuration.

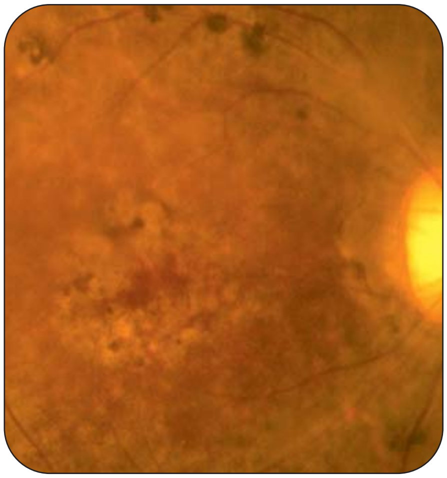

Fig. 13.157

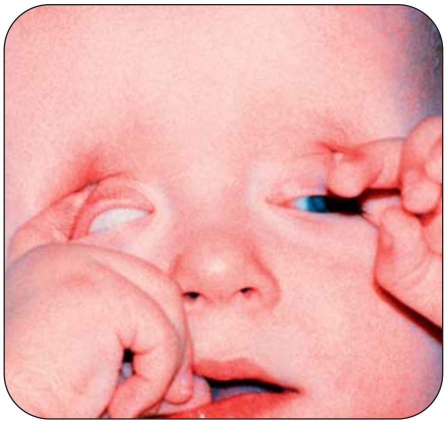

Fig. 13.158

Fibrotic lesions

Cellophane maculopathy

Signs

- Transparent membrane which may be associated with subtle retinal striae and vascular tortuosity.
- Best detected with red-free light (**Fig. 13.159**).

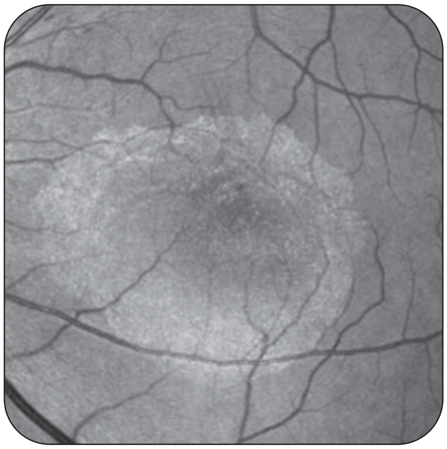

Fig. 13.159

Pucker

Signs

- An opaque membrane that frequently radiates from an epicentre and causes vascular distortion (**Fig. 13.160**).

Causes

- Idiopathic.
- Chronic intraocular inflammation.
- Retinal vascular disorders.
- Iatrogenic – scleral buckling procedures, retinal cryotherapy and retinal photocoagulation.
- Combined hamartoma of retina and RPE.

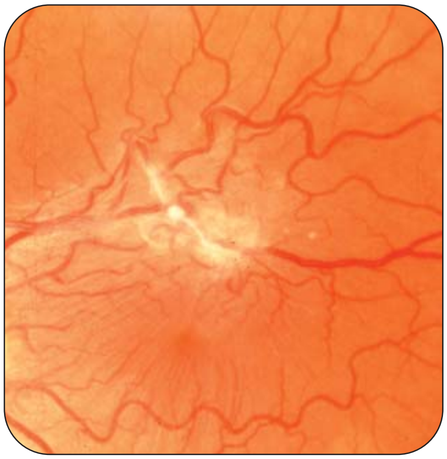

Fig. 13.160

Subretinal fibrosis (disciform scar)

Signs

- White-grey oval or round subretinal scar at the macula (**Fig. 13.161**) in eyes with end-stage of exudative maculopathy associated with CNV (see above).

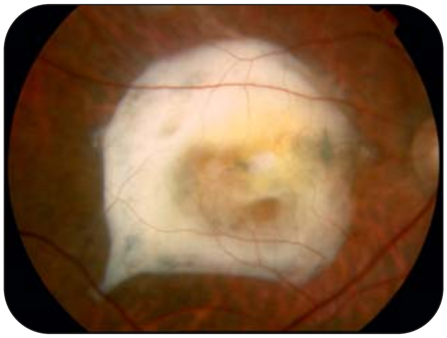

Fig. 13.161

Diffuse subretinal fibrosis

Signs

- Irregular greyish-white subretinal opacification often with distortion of the retinal vasculature (**Fig. 13.162**).

Causes

- Progressive subretinal fibrosis and uveitis syndrome.
- Multifocal choroiditis with panuveitis (**Fig. 13.163**) is an occasional cause.
- Vogt–Koyanagi–Harada (**Fig. 13.164**).

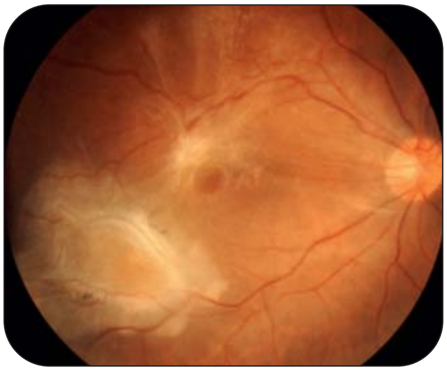

Fig. 13.162

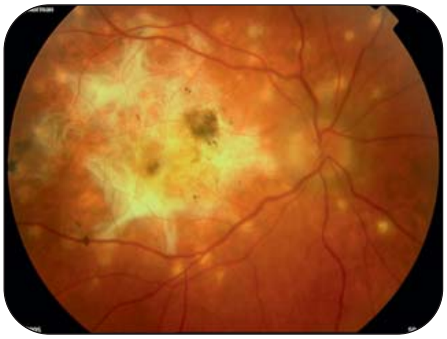

Fig. 13.163

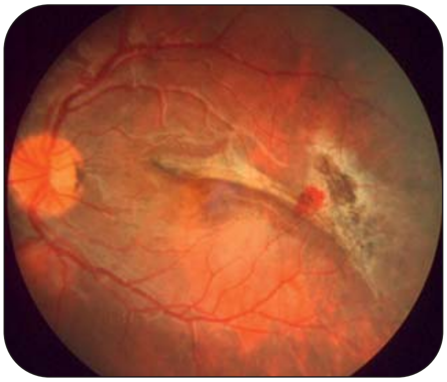

Fig. 13.164

Hereditary crystalline maculopathy

Primary oxalosis type 1

Signs

- Initially there are yellow crystals at the posterior pole.
- Later RPE hyperplasia develops (**Fig. 13.165**).

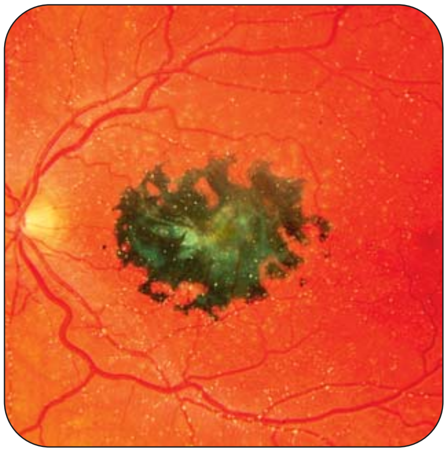

Fig. 13.165

Bietti crystalline dystrophy

Signs

- Initially there are fine, glistening, yellow-white crystals scattered throughout the posterior fundus (**Fig. 13.166**).
- Subsequently diffuse atrophy of the choriocapillaris ensues.

Look for

- Crystalline keratopathy (see Fig. 5.97) is common.

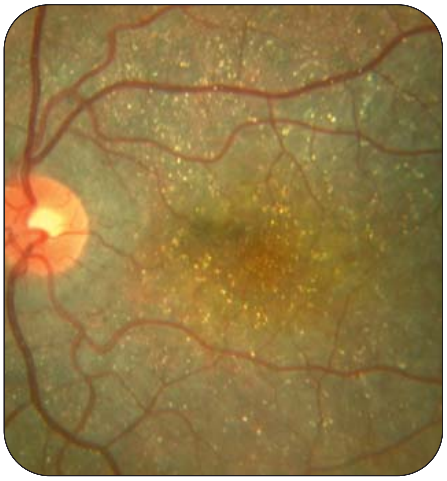

Fig. 13.166

AD crystalline retinopathy

Signs

- Similar to that of Bietti dystrophy but not associated with keratopathy.

Sjögren–Larsson syndrome

Signs

- Glistening yellow-white crystalline deposits at the macula present at birth (**Fig. 13.167**).

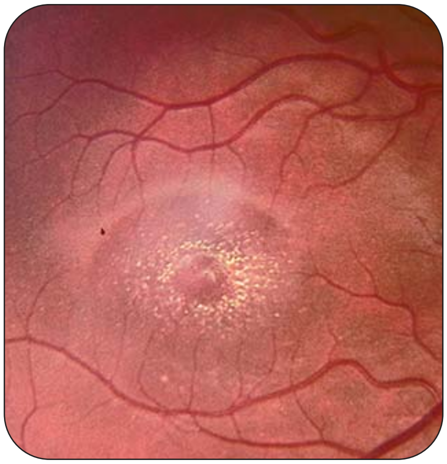

Fig. 13.167

Cystinosis

Signs

- Fine retinal crystals are uncommon.
- Corneal crystals are universal (see Fig. 5.90).

Drug-induced crystalline maculopathy

Tamoxifen

Indications

- To treat breast carcinoma; retinal toxicity is uncommon.

Signs

- Fine, superficial, yellow, crystalline deposits and deeper punctate grey lesions (**Fig. 13.168**).

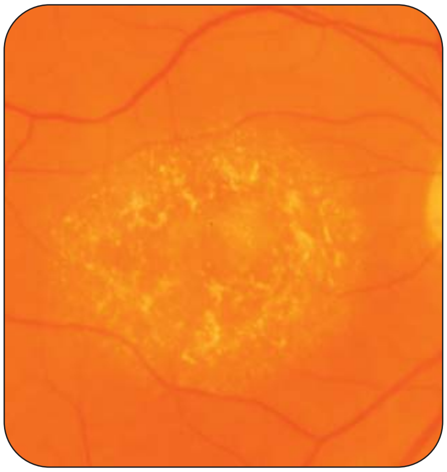

Fig. 13.168

Canthaxanthin

Indications

- To enhance sun tanning; retinal toxicity is uncommon and innocuous.

Signs

- Tiny, glistening, inner retinal yellow deposits, arranged symmetrically in a doughnut shape at the posterior pole (**Fig. 13.169**).

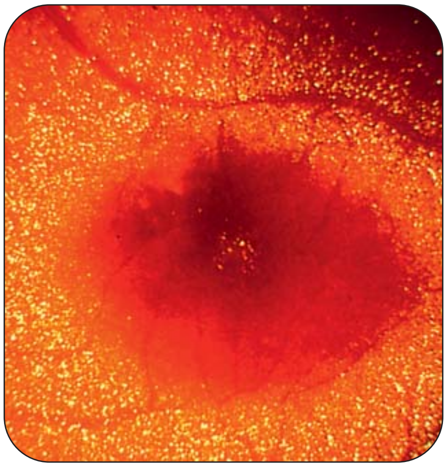

Fig. 13.169

Methoxyflurane

Indications

- An inhalant general anaesthetic, prolonged administration of which may lead to renal failure and secondary hyperoxalosis.

Signs

- The retinal changes are identical to those seen in primary oxalosis type 1 (see Fig. 13.165).

Miscellaneous crystalline maculopathy

Drusen

Signs

- Calcification within long-standing drusen (**Fig. 13.170**).

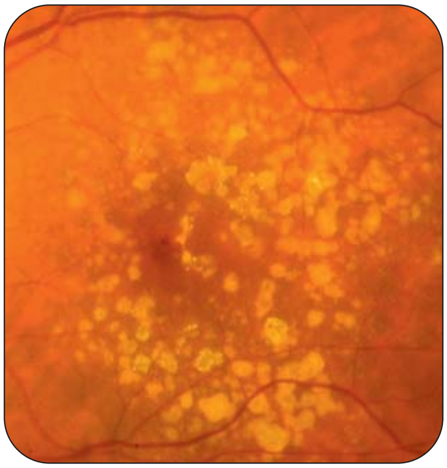

Fig. 13.170

Gyrate atrophy

Signs

- Subtle small glistening crystals are seen occasionally (**Fig. 13.171**).

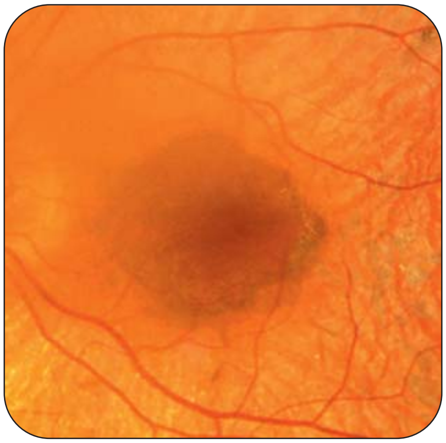

Fig. 13.171

2A bilateral idiopathic acquired parafoveolar telangiectasia

Signs

- Yellow crystalline deposits are common (**Fig. 13.172**).

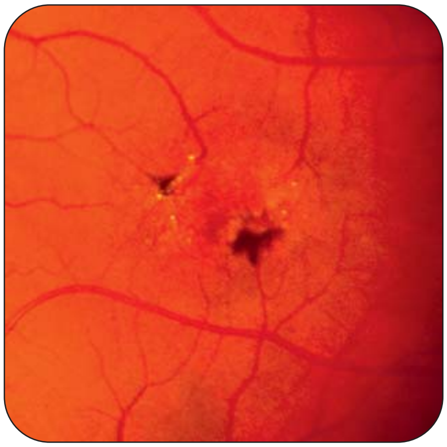

Fig. 13.172

Macular hole

Idiopathic (age-related)

Signs

- Central foveal defect with multiple yellow deposits within the crater surrounded by a cuff of subretinal fluid (**Fig. 13.173**).
- OCT shows a foveal defect that may be associated with a detached operculum (**Fig. 13.174**).

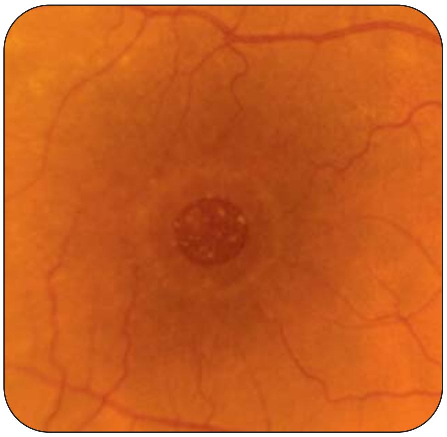

Fig. 13.173

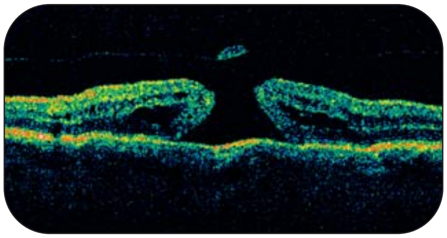

Fig. 13.174

Microhole

Signs

- Very small, red, well-demarcated foveal or juxtafoveal defect (**Fig. 13.175**).
- OCT shows a localised defect on the photoreceptors or RPE (**Fig. 13.176**).

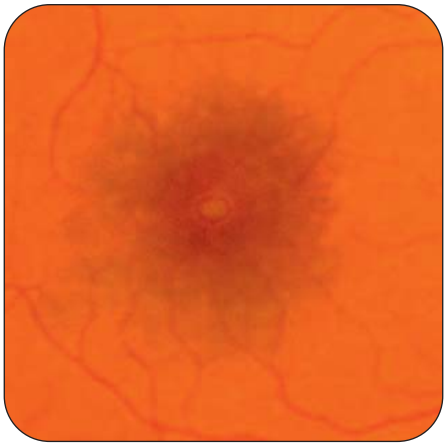

Fig. 13.175

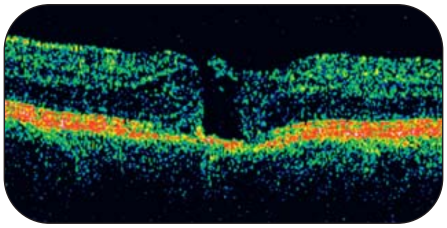

Fig. 13.176

Myopic

Unlike age-related macular hole it may give rise to retinal detachment which is mainly confined to the posterior pole (**Fig. 13.177**).

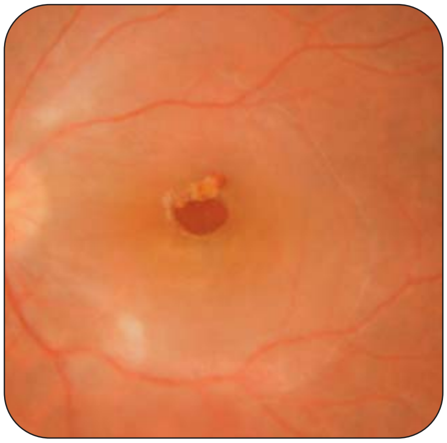

Fig. 13.177

Traumatic

Pathogenesis

- Severe Berlin oedema (commotio retinae – **Fig. 13.178**) may subsequently result in the formation of either an outer lamellar hole or a full-thickness macular hole.

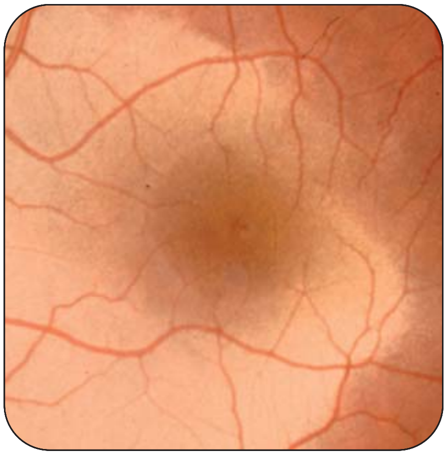

Fig. 13.178

Causes of macular pseudo-hole

- An inner lamellar hole caused by CMO (**Fig. 13.179**).
- A round discontinuity in an epiretinal membrane overlying the macula (**Fig. 13.180**). OCT confirms the presence of membrane and absence of a true hole (**Fig. 13.181**).
- In eyes with rhegmatogenous retinal detachment involving the posterior pole, a macular pseudo-hole may be seen because of the thinness of the retina at the fovea (**Fig. 13.182**).

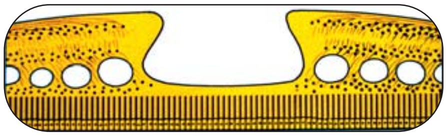

Fig. 13.179

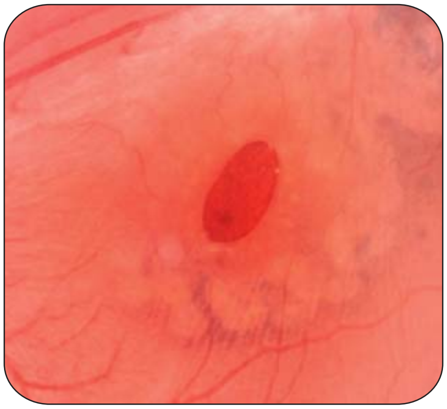

Fig. 13.180

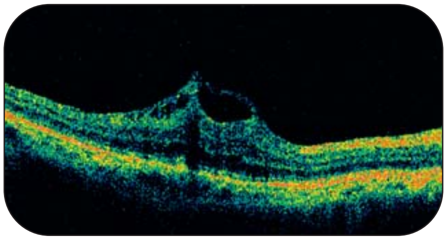

Fig. 13.181

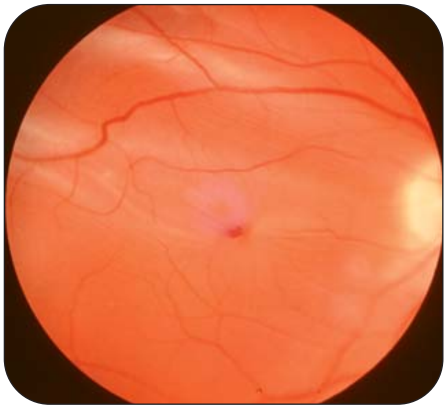

Fig. 13.182

PERIPHERAL RETINAL DEGENERATIONS

Innocuous

Honeycomb (reticular)

Signs

- Bilateral, fine network of pigmentation that may extend posterior to the equator (**Fig. 13.183**).

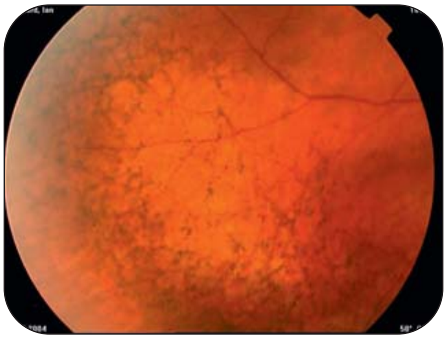
Fig. 13.183

Peripheral drusen

Signs

- Multitude of tiny pale dots that may be associated with mild pigmentary changes (**Fig. 13.184**).

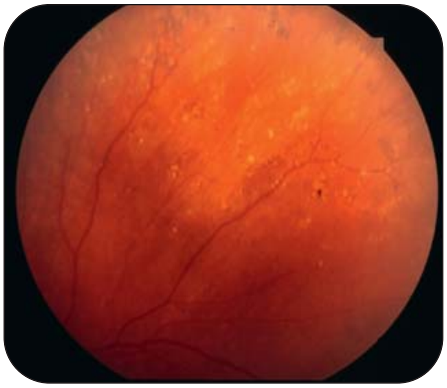
Fig. 13.184

Microcystoid

Signs

- Tiny vesicles with indistinct borders on a greyish-white background (**Fig. 13.185**).

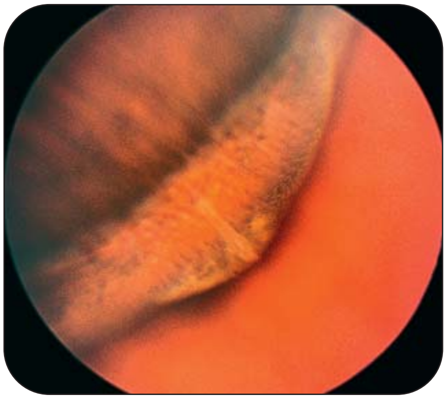
Fig. 13.185

Pavingstone

Signs

- Discrete, yellow-white patches of focal chorioretinal atrophy (**Fig. 13.186**).

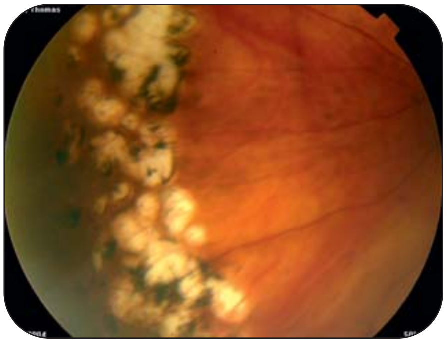

Fig. 13.186

Predisposing

Lattice

Prevalence

- Present in 8% of the general population and in 40% of eyes with RD.

Signs

- Spindle shaped areas containing an arborising network of white lines and variable RPE hyperplasia (**Fig. 13.187**).

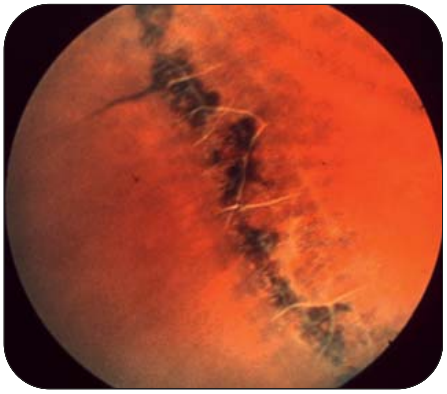

Fig. 13.187

Snailtrack

Prevalence

- An uncommon condition that is occasionally associated with RD.

Signs

- Sharply demarcated bands of tightly packed snowflakes (**Fig. 13.188**).

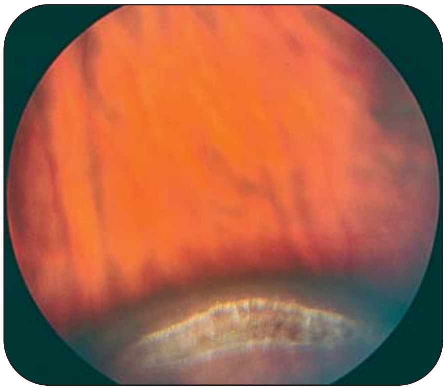

Fig. 13.188

White-without-pressure

Prevalence

- An uncommon condition that is occasionally associated with giant retinal tear formation.

Signs

- Superficial grey area with a geographic configuration (**Fig. 13.189**).

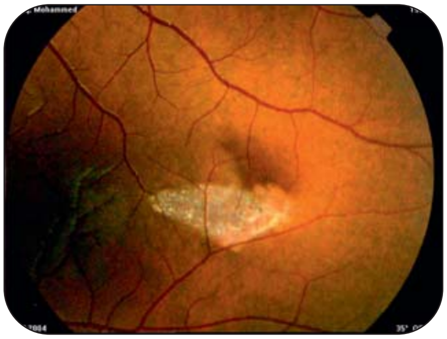

Fig. 13.189

RETINAL DETACHMENT

Rhegmatogenous

Rhegmatogenous retinal detachment (RD) is caused by a retinal break which may be a hole or a tear.

Fresh

Signs

- The detached retina has a slightly opaque and corrugated appearance, and blood vessels appear darker than in attached retina (**Fig. 13.190**).
- The subretinal fluid (SRF) has a convex configuration (**Fig. 13.191**) and retinal breaks appear as discontinuities of the retinal surface (**Fig. 13.192**).
- Ultrasonography shows a mobile membrane attached anteriorly to the ora serrata and posteriorly to the optic nerve head (**Fig. 13.193**).

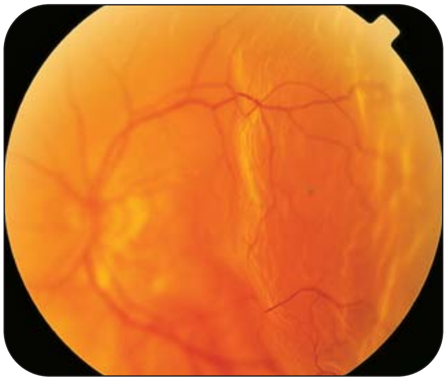

Fig. 13.190

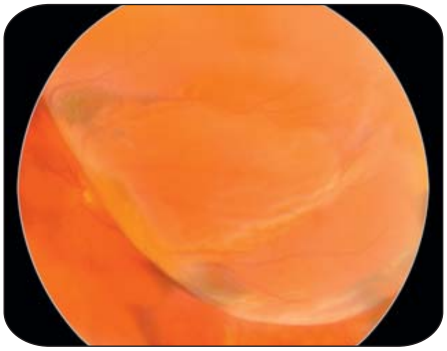

Fig. 13.191

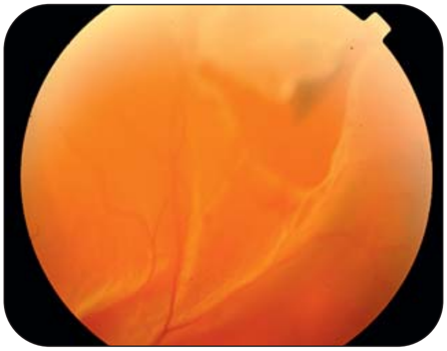

Fig. 13.192

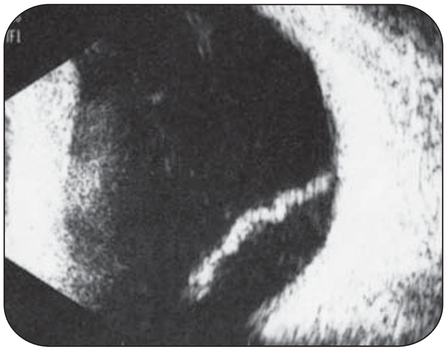

Fig. 13.193

Proliferative vitreoretinopathy

Signs

- Retinal stiffness and rigid folds (**Fig. 13.194**).

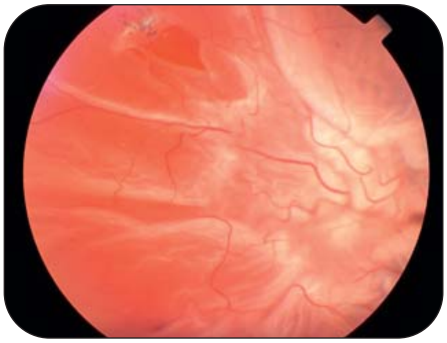

Fig. 13.194

Long-standing

Signs

- Thin retina which contains secondary intraretinal cysts (**Fig. 13.195** and **Fig. 13.196**).
- Demarcation lines (high water marks) at the junction of attached and detached retina (**Fig. 13.197**).

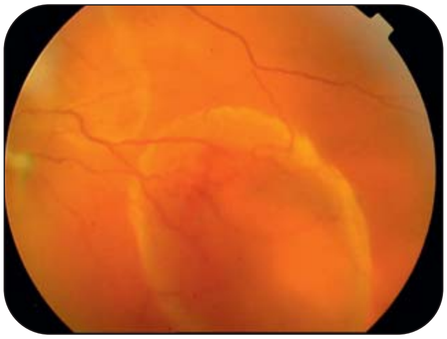

Fig. 13.195

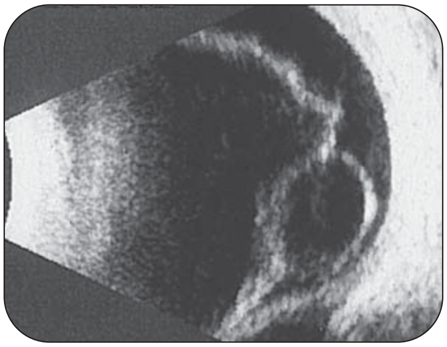

Fig. 13.196

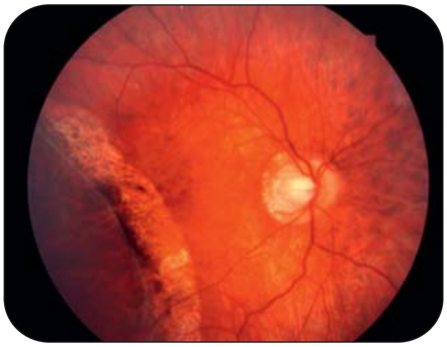

Fig. 13.197

Tractional

Causes

- Proliferative diabetic retinopathy and penetrating trauma are the most common.

Signs

- SRF is concave and retinal breaks absent (**Fig. 13.198**).
- Ultrasonography shows a relatively immobile membrane; diabetic detachments may have coexisting vitreous blood (**Fig. 13.199**).

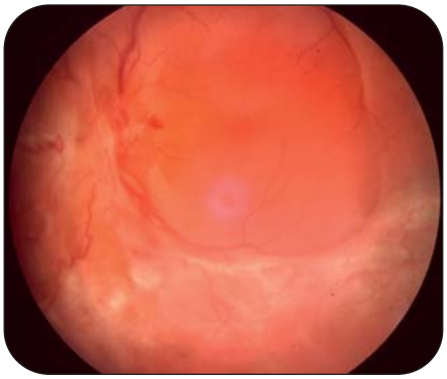

Fig. 13.198

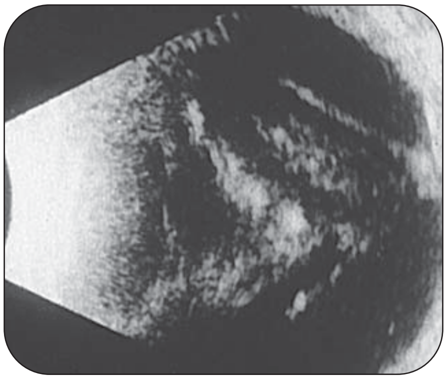

Fig. 13.199

Exudative

Causes

- See Table 13.5.

Signs

- The detached retina has a smooth surface with absence of retinal breaks; its cause (such as a choroidal tumour) may be evident (**Fig. 13.200**).
- SRF is very mobile and deep, and extends to the ora serrata.
- Scattered subretinal pigment clumps ('leopard spots') may be seen when the retina flattens (see Fig. 13.208).

Table 13.5 **Causes of exudative RD**

1. **Choroidal tumours**
    - Primary
    - Metastatic
2. **Systemic**
    - Hypertension
    - Toxaemia of pregnancy
    - Hypoproteinaemia
3. **Intraocular inflammation**
    - Harada disease
    - Posterior scleritis
    - Necrotising retinitis
4. **Leaking subretinal vessels**
    - Choroidal neovascularisation
    - Retinal telangiectasia
5. **Iatrogenic**
    - Retinal detachment surgery
    - Extensive retinal photocoagulation

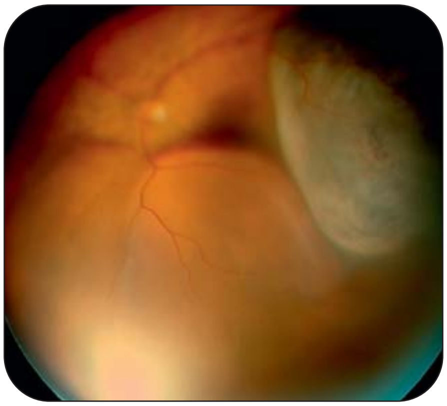

Fig. 13.200

Differential diagnosis

Degenerative retinoschisis

Signs

- Smooth, very thin, relatively immobile retinal elevation in which the inner layer may show 'snowflakes' and vascular sheathing (**Fig. 13.201**).
- Breaks may be absent, or present in one (**Fig. 13.202**) or both layers.

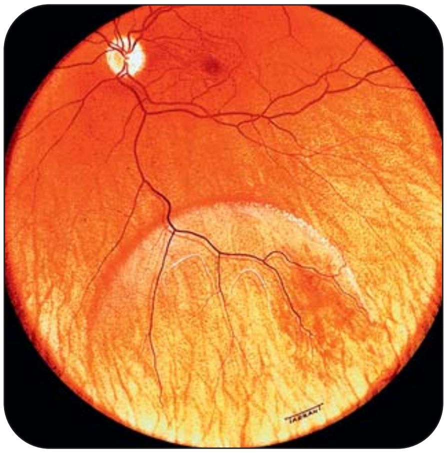

Fig. 13.201

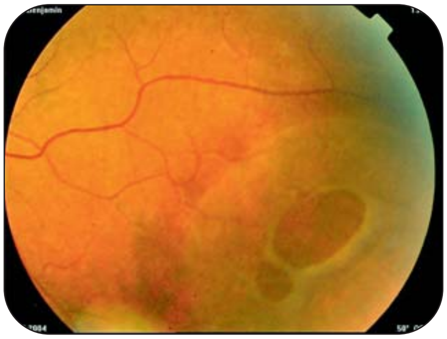

Fig. 13.202

Choroidal detachment

Signs

- Brown, convex, smooth, peripheral elevations (**Fig. 13.203**).
- Very low intraocular pressure.
- Ultrasonography shows immobile dome-shaped elevations which may almost touch ('kissing choroidals' – **Fig. 13.204**).

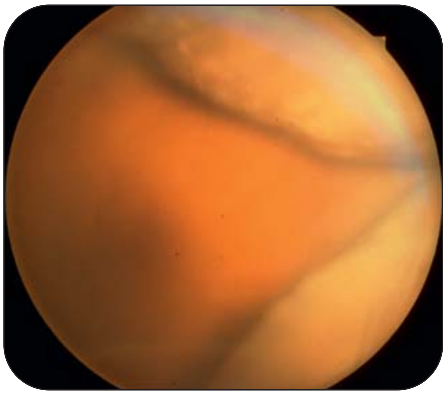

Fig. 13.203

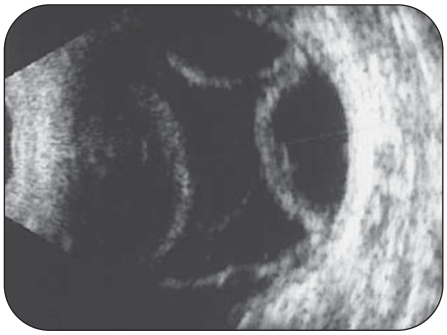

Fig. 13.204

Uveal effusion syndrome

Definition

- A rare usually idiopathic condition that may be bilateral.

Signs

- Ciliochoroidal detachment associated with exudative retinal detachment (**Fig. 13.205**).

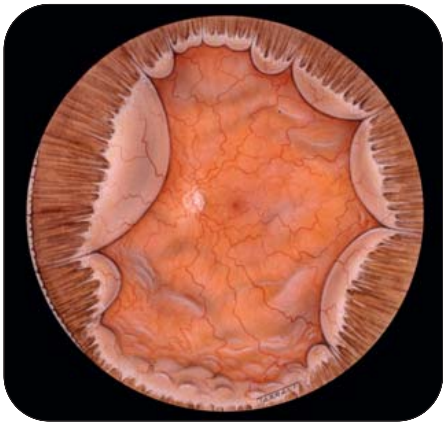

Fig. 13.205

SCATTERED PIGMENTARY CHANGES

Clumping

Chloroquine retinotoxicity

Toxicity may occur when large doses have been administered for a long time.

Signs

- Bull's eye maculopathy occurs in advanced cases (see Fig. 13.155).
- Pigment clumping, that may be perivascular, and atrophy of the RPE occurs in end-stage disease (**Fig. 13.206**).

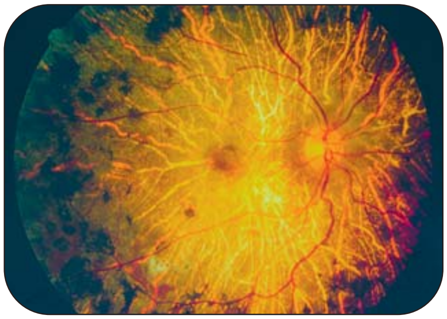

Fig. 13.206

Thioridazine retinotoxicity

Toxicity may occur within a few weeks if excessive doses are administered.

Signs

- Pigment clumps and plaques with loss of RPE occur in end-stage disease (**Fig. 13.207**).

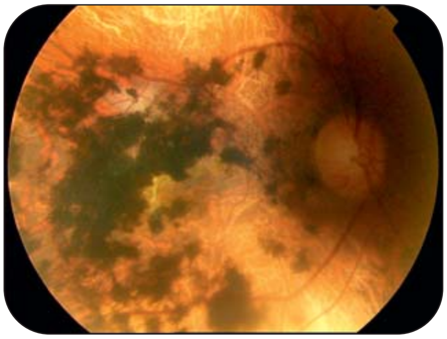

Fig. 13.207

'Leopard spots'

This appearance may develop following resolution of exudative RD.

Signs

- Scattered subretinal pigment clumps (**Fig. 13.208**).

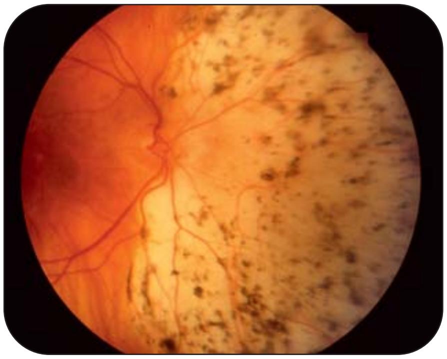

Fig. 13.208

Grouped congenital hypertrophy of the RPE

Signs

- Small, circumscribed, oval black spots, often organised in a pattern simulating animal footprints (**Fig. 13.209**).

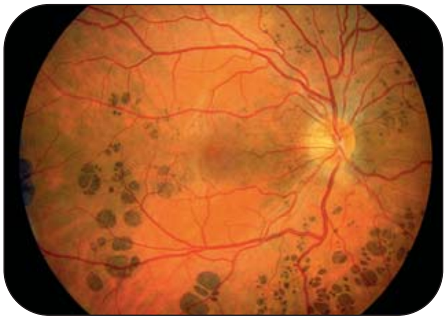

Fig. 13.209

Retinitis pigmentosa

Signs

a. *Typical* (rod-cone dystrophy) – bone-spicule perivascular pigmentation, arteriolar attenuation and waxy disc pallor (**Fig. 13.210**).

b. *Inverse* (cone-rod dystrophy) – confined to the posterior pole (**Fig. 13.211**).

c. *Sectoral* – confined to only one quadrant (**Fig. 13.212**) or one half.

Systemic associations

- see Table 13.6.

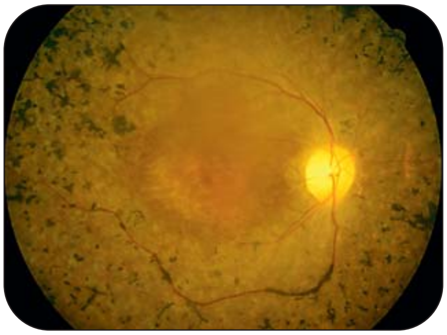

Fig. 13.210

Table 13.6 **Systemic associations of retinitis pigmentosa**

- Refsum disease
- Bassen–Kornzweig syndrome
- Usher syndrome (types I and II)
- Laurence–Moon syndrome
- Bardet–Biedl syndrome
- Kearns–Sayre syndrome
- Cockayne syndrome
- Alström syndrome
- Olivopontocerebellar atrophy
- Batten–Vogt disease
- Hallervorden–Spatz syndrome
- Mucopolysaccharidosis (types I and II)
- Mucolipidosis type IV
- Jeune syndrome
- Sly syndrome

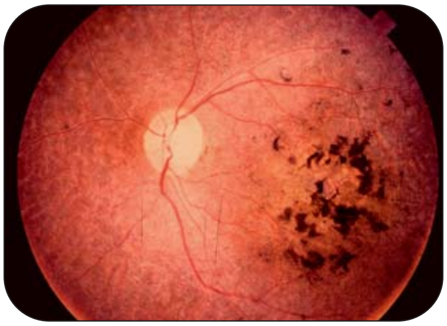

Fig. 13.211

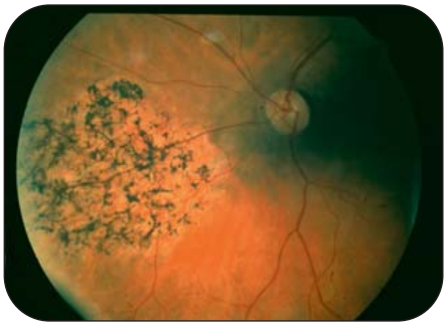

Fig. 13.212

End-stage acquired syphilitic chorioretinitis

Signs

- Vascular attenuation optic atrophy and pigment migration (**Fig. 13.213**).

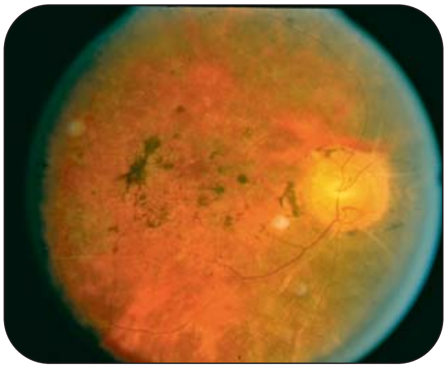

Fig. 13.213

Pigmented paravenous chorioretinal atrophy

Definition

- An innocuous asymptomatic condition of uncertain inheritance.

Signs

- Perivenous bone-spicule pigmentation associated with chorioretinal atrophy (**Fig. 13.214**).

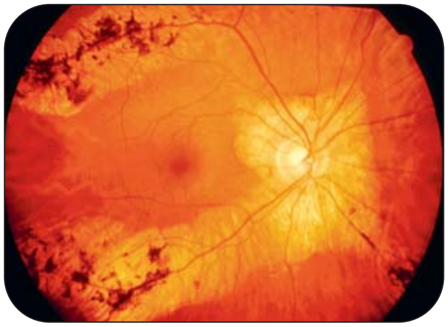

Fig. 13.214

Enhanced S-cone syndrome

Definition

- An AR dystrophy that presents with nyctalopia in childhood.

Signs

- Peripheral pigment clumping (**Fig. 13.215**) and macular atrophy.

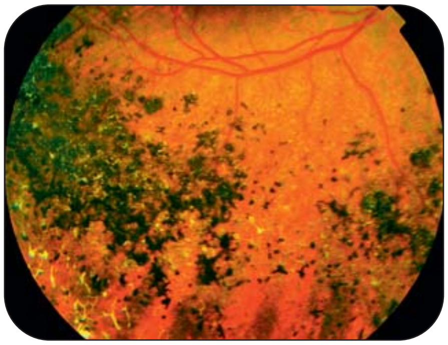

Fig. 13.215

Mottling

Rubella retinopathy

Definition

- A common and usually innocuous manifestation of congenital rubella.

Signs

- Generalised mottled pigmentation most marked at the posterior pole (**Fig. 13.216**).

Complications

- CNV is rare.

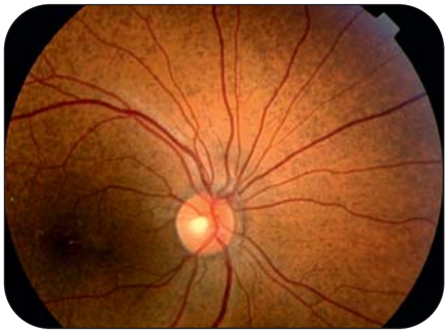

Fig. 13.216

Congenital syphilis

Signs

- Salt and pepper fundus (**Fig. 13.217**) with normal vision in mild cases.
- Pigmentary retinopathy resembling retinitis pigmentosa is uncommon.

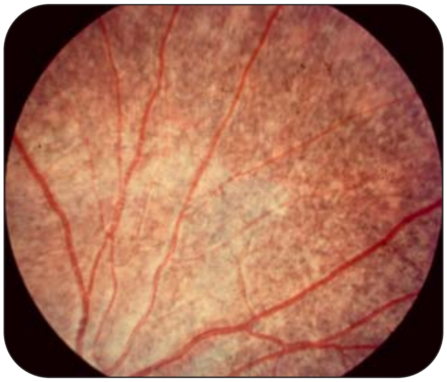

Fig. 13.217

'Peau d'orange'

Signs

- Mottled pigmentation of the posterior fundus in eyes with angioid streaks (**Fig. 13.218**).
- In some eyes the appearance is striking and subtle angioid streaks may be overlooked, in other cases 'peau d'orange' is subtle or non-existent.

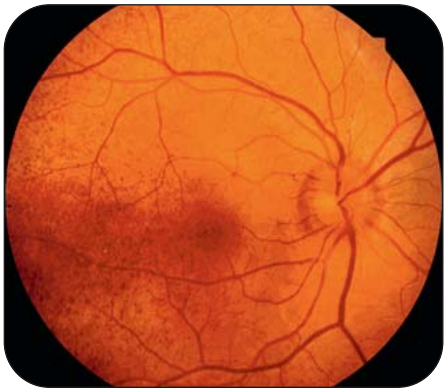

Fig. 13.218

Female carriers of choroideremia

Signs

- Patchy peripheral atrophy and mottling of the RPE (**Fig. 13.219**).

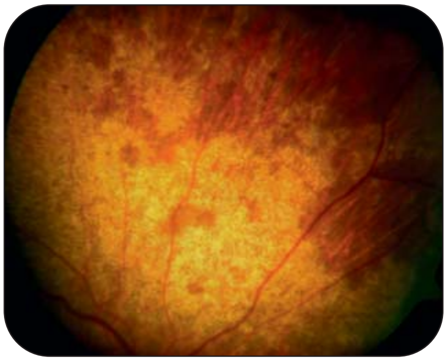

Fig. 13.219

MULTIPLE YELLOW-WHITE SPOTS AND FLECKS

Hard drusen

Definition

- Very common, bilateral, innocuous age-related changes.

Signs

- Multiple, round, discrete, yellow-white lesions less than half a vein width in diameter with well-defined borders (**Fig. 13.220**).

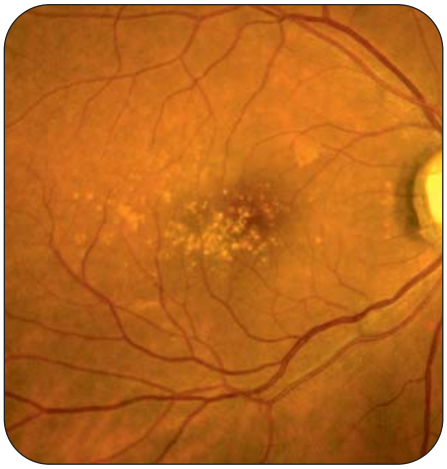

Fig. 13.220

Soft drusen

Definition

- Bilateral age-related changes that carry an increased risk of AMD.

Signs

- Larger than hard drusen with poorly delineated margins (**Fig. 13.221**).

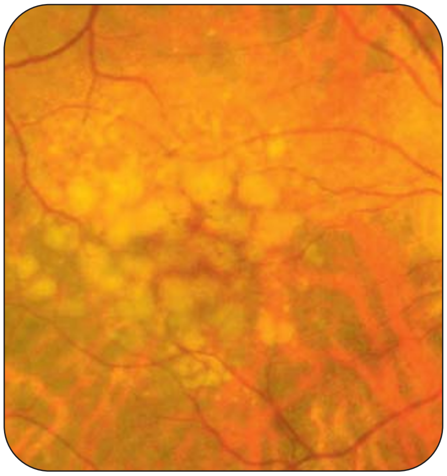

Fig. 13.221

Cuticular (basal laminar) drusen

Definition

- Uncommon bilateral condition that occurs in middle-aged individuals and is associated with an increased risk of AMD.

Signs

- Innumerable small discrete, round, slightly raised, yellow subretinal lesions (**Fig. 13.222**).
- FA shows hyperfluorescent spots reminiscent of a starry-night (Milky-Way appearance – **Fig. 13.223**).

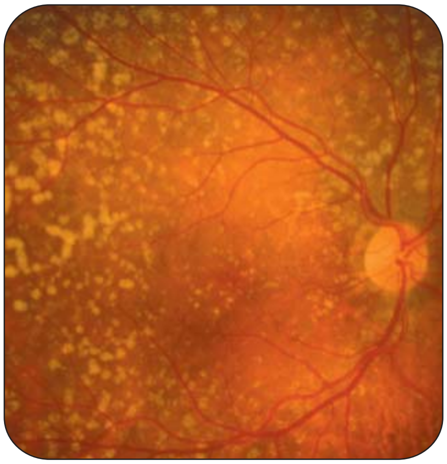

Fig. 13.222

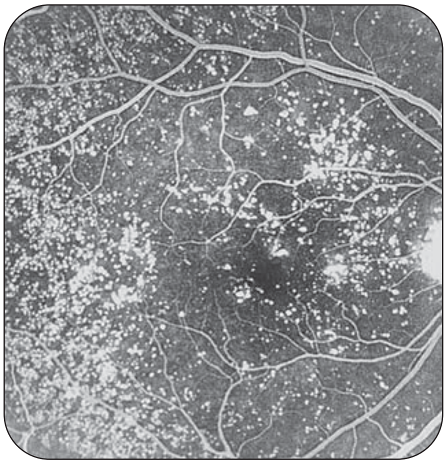

Fig. 13.223

Familial drusen (Doyne honeycomb dystrophy)

Definition

- An uncommon AD condition which appears during the late teens or early twenties and may later be associated with AMD.

Signs

- Soft drusen often with a symmetrical distribution in both eyes such as temporal to the fovea or nasal to the disc (**Fig. 13.224**).

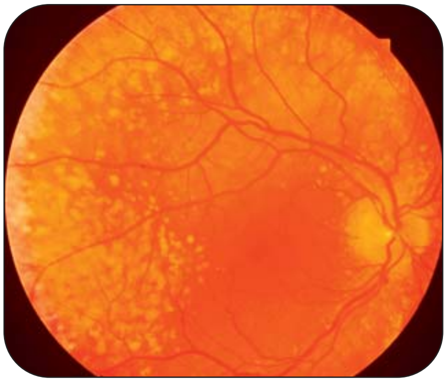

Fig. 13.224

Malattia levantinese

Definition

- A very rare AD condition that shares phenotypic overlap with familial drusen.

Signs

- Diffuse, yellowish, round spots of variable size arranged in a spoke-like or radial pattern.
- Later the spots tend to merge, forming a white circle with indistinct edges (**Fig. 13.225**).

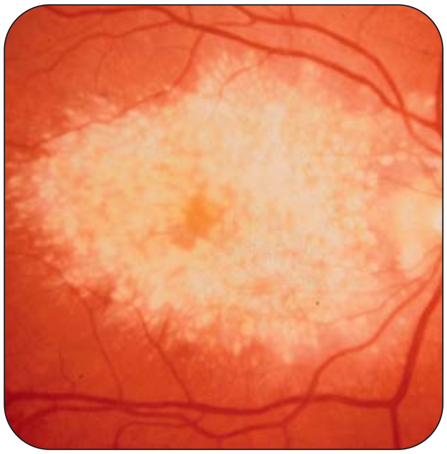

Fig. 13.225

Fundus flavimaculatus

Definition

- An AR condition, which in the absence of macular involvement, may remain asymptomatic throughout life.

Signs

- Yellow-white, fish-tail-shaped flecks at the posterior pole and midperiphery (**Fig. 13.226**).
- The flecks may occur in isolation or in association with maculopathy (Stargardt macular dystrophy – see Figs 13.151, 13.152 and 13.153).
- FA in long-standing cases shows hyperfluorescence due to RPE atrophy. A dark choroid is seen in 85% of cases (**Fig. 13.227**).

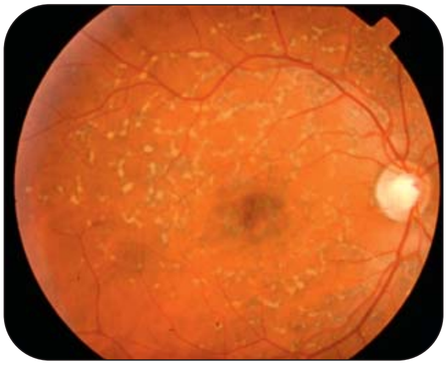

Fig. 13.226

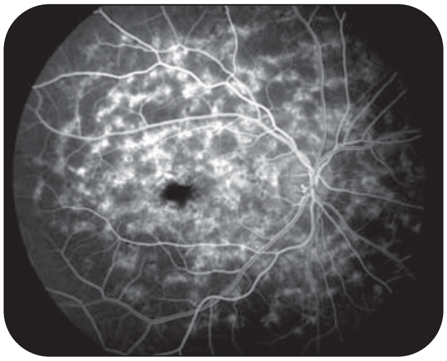

Fig. 13.227

Fundus albipunctatus

Definition

- An AR condition characterised by congenital non-progressive night-blindness.

Signs

- A multitude of tiny yellow-white spots extends from the posterior pole, where they are most numerous, to the periphery (**Fig. 13.228**).

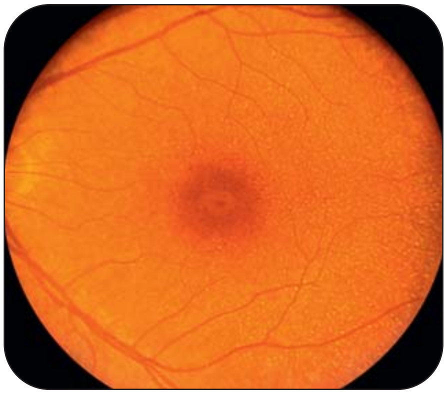

Fig. 13.228

Retinitis punctata albescens

Definition

- A rare subtype of RP.

Signs

- Discrete white dots and arteriolar attenuation (**Fig. 13.229**).
- FA shows mottled hyperfluorescence (**Fig. 13.230**).

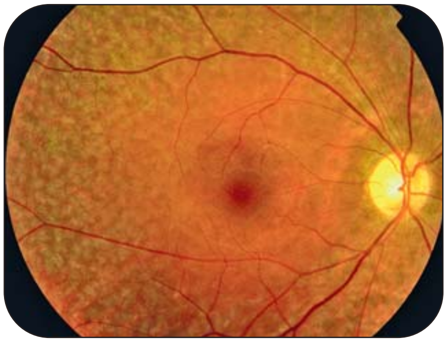

Fig. 13.229

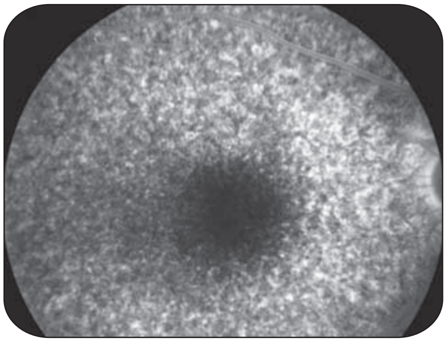

Fig. 13.230

Benign flecked retina

Definition

- An innocuous AR condition.

Signs

- Widespread, discrete, yellow-white polymorphous flecks that spare the macula (**Fig. 13.231**).

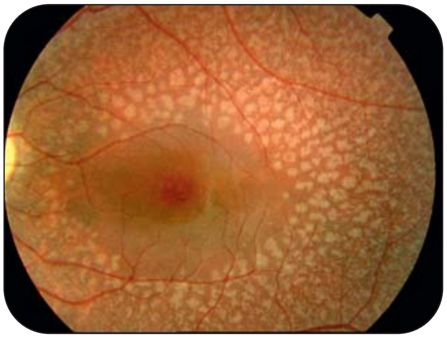

Fig. 13.231

Alport syndrome

Definition

- An XL disease characterised by progressive renal disease and deafness.

Signs

- Pale punctate yellow flecks at the posterior pole (**Fig. 13.232**).
- Confluent and clustered yellow flecks in the midperiphery (**Fig. 13.233**).

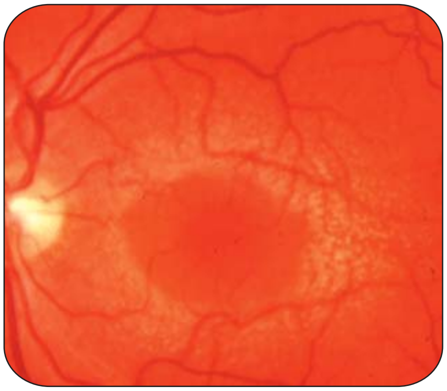

Fig. 13.232

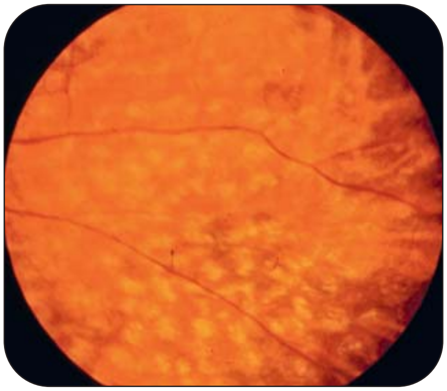

Fig. 13.233

Look for

- Anterior lenticonus and microspherophakia.

North Carolina macular dystrophy

Definition

- A very rare AD condition with variable manifestations.

Signs

- Yellow-white drusen-like deposits at the macula (**Fig. 13.234**) and in the periphery (**Fig. 13.235**).

Complications

- CNV may develop.

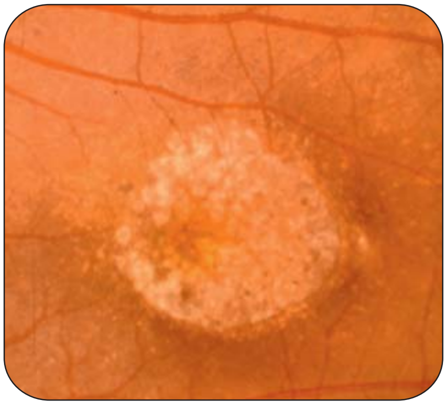

Fig. 13.234

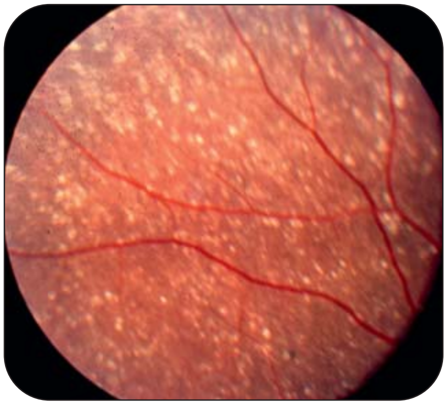

Fig. 13.235

Sorsby pseudo-inflammatory dystrophy

Definition

- A very rare AD condition that usually causes severe visual loss.

Signs

- Yellow-white confluent deposits along the vascular arcades, nasal to the disc and mid-periphery (**Fig. 13.236**).

Complications

- CNV is very common.

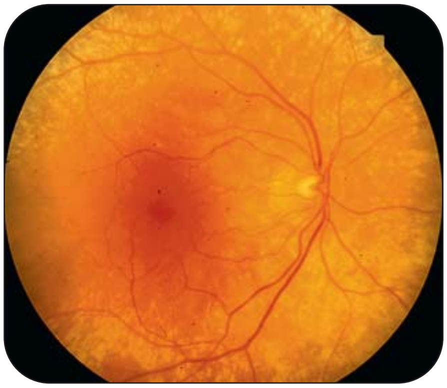

Fig. 13.236

FLAT FOCAL LESIONS

Black

Typical congenital RPE hypertrophy

Definition

- A very common innocuous condition without systemic implications.

Signs

- Usually solitary, circular, dark-grey or black, placoid lesion, one to two disc diameters in size, with smooth or scalloped borders (**Fig. 13.237**).
- Occasionally the lesion may contain hypopigmented lacunae (**Fig. 13.238**).

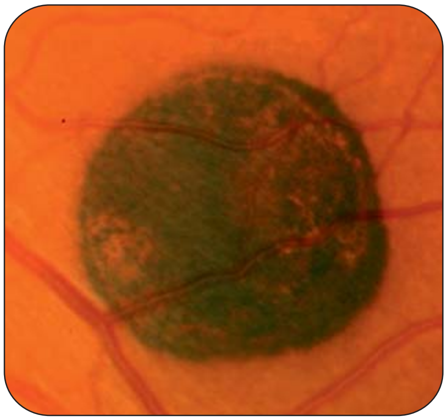

Fig. 13.237

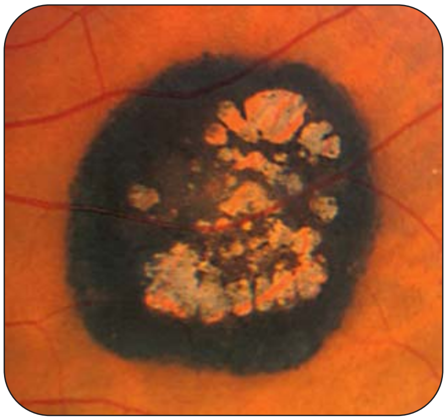

Fig. 13.238

Atypical congenital RPE hypertrophy

Definition

- A rare condition that may be associated with familial adenomatous polyposis, Gardner syndrome and Turcot syndrome.

Signs

- Multiple, bilateral, oval or spindle-shaped lesions with a hypopigmented outline at one margin (**Fig. 13.239**).

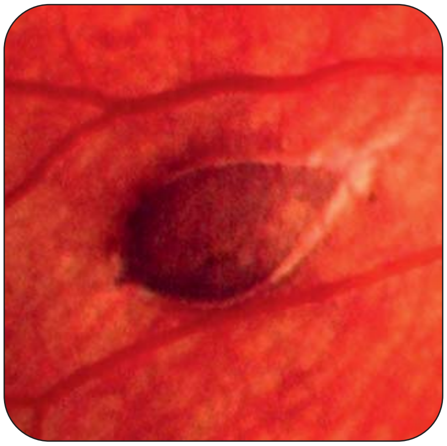

Fig. 13.239

Choroidal naevus

Signs

- Flat or minimally elevated, circular, slate-grey lesion <5 mm in diameter (**Fig. 13.240**).
- Surface drusen are common in long-standing lesions (**Fig. 13.241**).

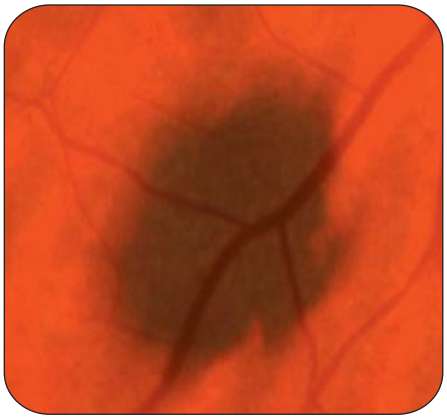

Fig. 13.240

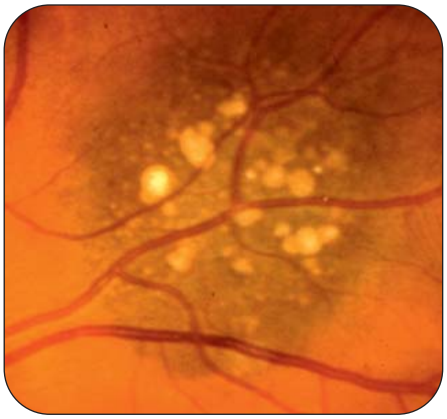

Fig. 13.241

RPE hamartoma

Signs

- Jet-black lesion at the macula that tends to spill onto the inner retinal surface (**Fig. 13.242**).

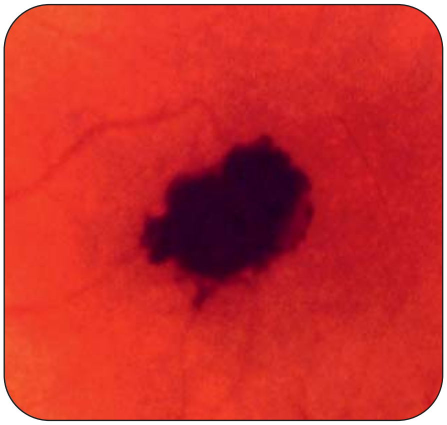

Fig. 13.242

Bilateral diffuse uveal melanocytic proliferation

Definition

- A very rare paraneoplastic syndrome usually occurring in patients with systemic malignancy which is often occult.

Signs

- Multiple naevus-like choroidal lesions (**Fig. 13.243**).
- Multiple red-grey subretinal patches and exudative RD.

Look for

- Cataract.
- Uveitis.
- Episcleral nodules.
- Anterior uveal cysts and tumours.

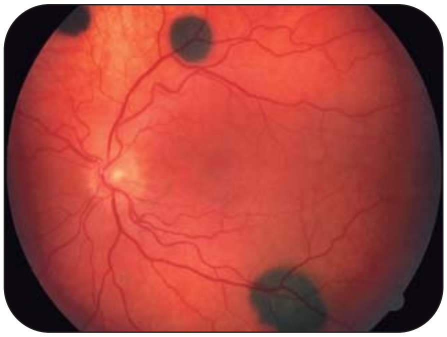

Fig. 13.243

Pale

Old retinochoroiditis

Signs

- Round or oval atrophic lesion with visible choroidal vessels and variable surrounding pigmentation (**Fig. 13.244**).

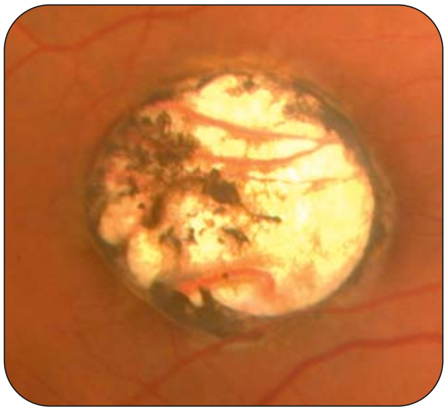

Fig. 13.244

Idiopathic chorioretinal atrophy

Signs

- Similar to old choroiditis but usually with less surrounding pigmentation (**Fig. 13.245**).
- Multiple small lesions in the peripheral fundus are referred to as 'paving-stone' degeneration (see Fig. 13.186).

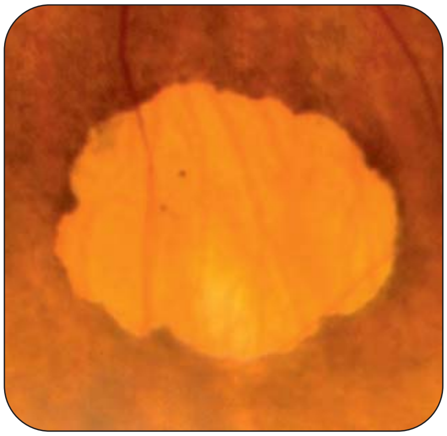

Fig. 13.245

Congenital hypertrophy of the RPE

Signs

- Solitary, circular, placoid lesion, one to two disc diameters in size, with smooth or scalloped borders in which most of the pigment has been absorbed (**Fig. 13.246**).

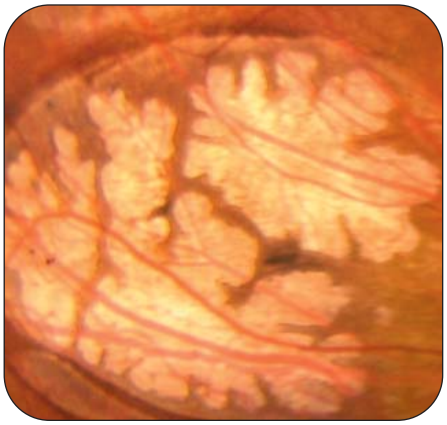

Fig. 13.246

Coloboma of the retinal and choroid

Signs

- Sharply circumscribed white area inferior to the disc largely devoid of blood vessels (**Fig. 13.247**).

Look for

- Other colobomas.

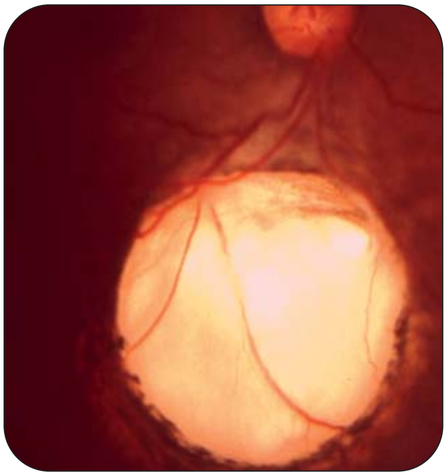

Fig. 13.247

Congenital albinotic spots

Signs

- Small, pale, circular lesions without pigmented outlines which usually occur in the peripheral fundus and rarely at the macula.
- Similar grouped spots are referred to as ‘polar-bear tracks’ (**Fig. 13.248**).

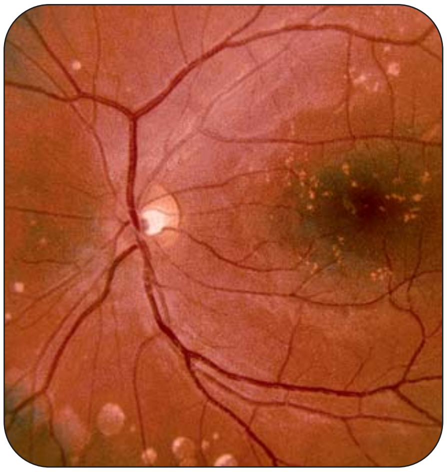

Fig. 13.248

Egg-yolk

Best vitelliform macular dystrophy

Definition

- A very rare AD condition which starts in childhood and is characterised by a subnormal EOG.

Signs

- Sunny-side up egg-yolk lesion (**Fig. 13.249**)
- FA shows hypofluorescence due to blockage (**Fig. 13.250**).

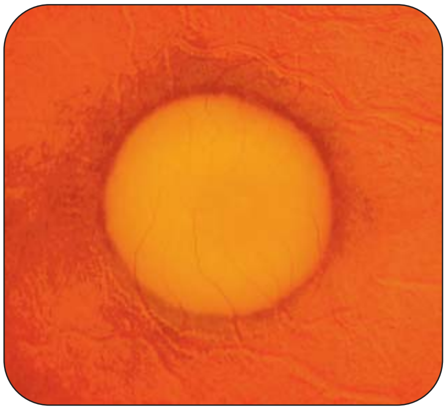
Fig. 13.249

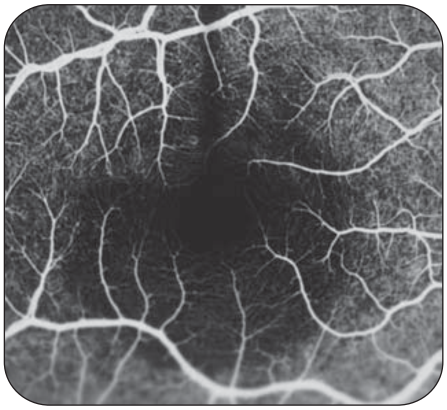
Fig. 13.250

Adult vitelliform macular dystrophy

Definition

- An uncommon relatively innocuous condition.

Signs

- Small, round, slightly elevated, yellow lesions at the fovea (**Fig. 13.251**).
- FA shows central hypofluorescence with a hyperfluorescent halo (**Fig. 13.252**).

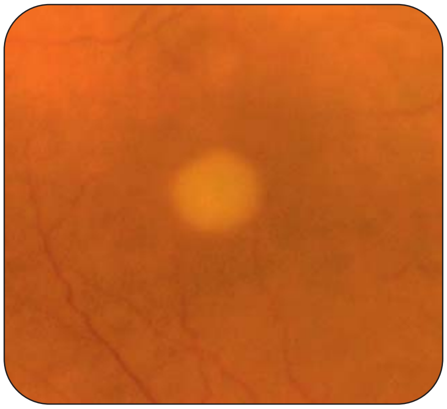
Fig. 13.251

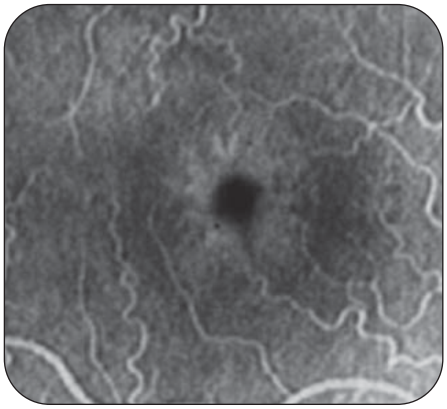
Fig. 13.252

Multifocal vitelliform lesions

Signs

- Multifocal lesions (**Fig. 13.253**) may occasionally occur in adults with no family history of Best disease and normal EOG.

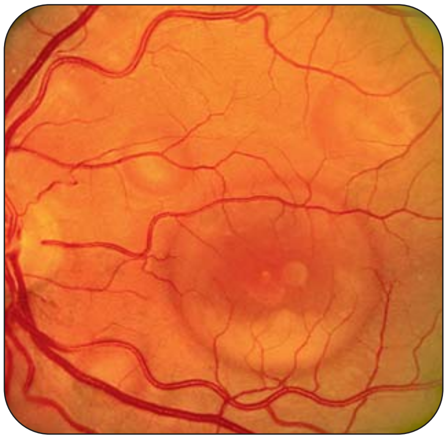
Fig. 13.253

Cherry-red spot

Acute central retinal artery occlusion

Signs

- The orange reflex from intact choroid stands out against the thin fovea in contrast to the surrounding pale ischaemic retina resulting from intracellular oedema (**Fig. 13.254**).
- Attenuated arteries with segmentation of the blood column ('cattle-tracking') in veins and arteries (**Fig. 13.255**).

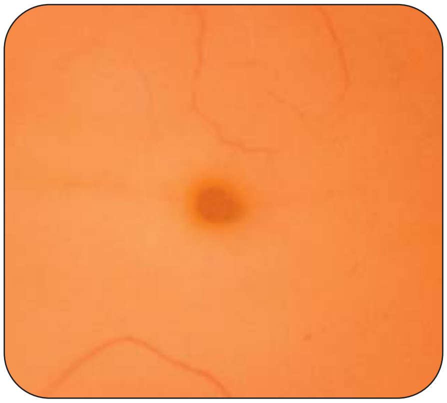

Fig. 13.254

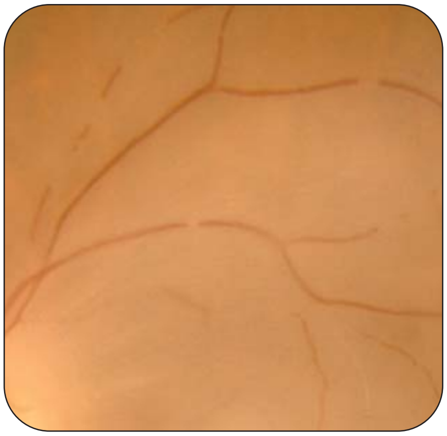

Fig. 13.255

Sphingolipidoses

Definition

- The sphingolipidoses encompass a group of rare metabolic disorders (e.g. Tay–Sachs disease and Niemann–Pick disease) characterised by relentless neurodegeneration and deposition of abnormal material in retinal ganglion cells that results in loss of transparency.

Signs

- The fovea retains its normal appearance because it is devoid of ganglion cells (**Fig. 13.256**).

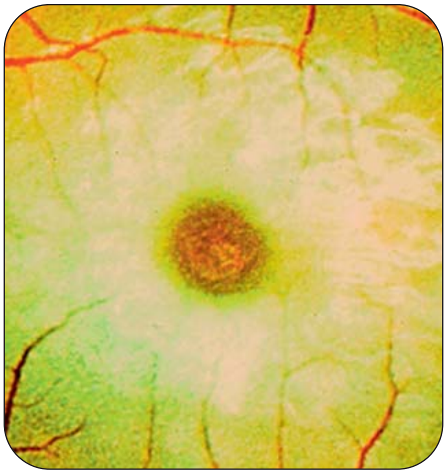

Fig. 13.256

FOCAL ELEVATED LESIONS

Retinal

Retinal astrocytoma

Definition

- A benign tumour that may be solitary or multifocal as well as bilateral. It is present in 50% of patients with tuberous sclerosis.

Signs

- Semi-translucent mulberry-like lesion (**Fig. 13.257**).
- FA shows hyperfluorescence due to staining but lack of leakage (**Fig. 13.258**).

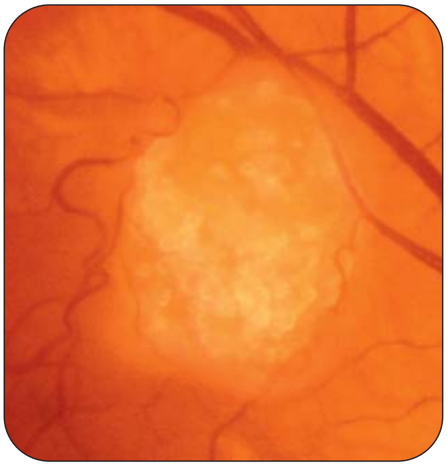

Fig. 13.257

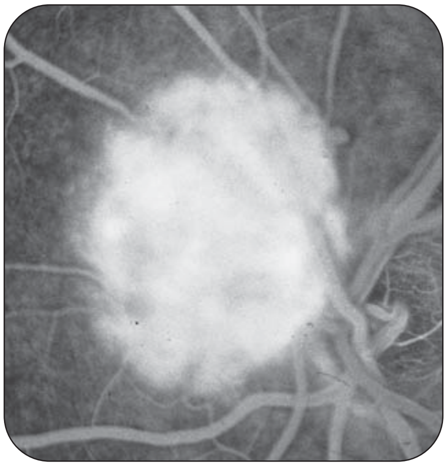

Fig. 13.258

Retinoblastoma

Definition

- The most common primary ocular malignancy in childhood that frequently presents with leukocoria (see Fig. 9.28).

Signs

- An endophytic tumour grows into the vitreous as a white mass (**Fig. 13.259**).
- Heritable tumours are usually bilateral and occasionally multifocal.
- Ultrasonography is useful in assessing tumour size and detecting calcification (**Fig. 13.260**).

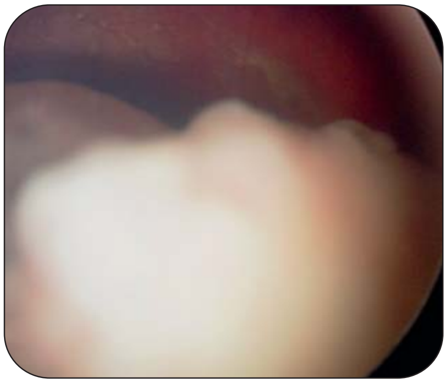

Fig. 13.259

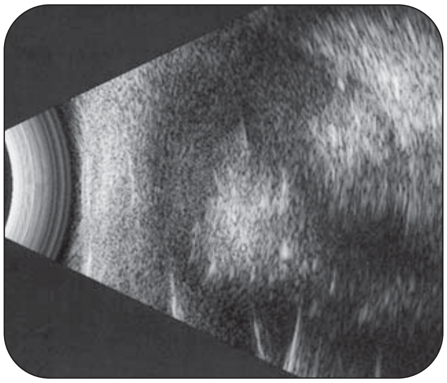

Fig. 13.260

Retinoma

Definition

- A very rare benign variant of retinoblastoma.

Signs

- White mass associated with RPE changes and chorioretinal atrophy (**Fig. 13.261**).

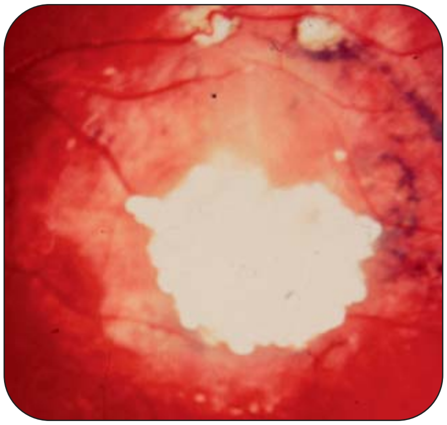

Fig. 13.261

Toxocara granuloma

Signs

- Always unilateral, round, yellow-white lesion in the posterior fundus (**Fig. 13.262**).
- Occasionally surrounded by hard exudates.

Complications

- Vitreoretinal traction bands.
- Localised tractional RD.

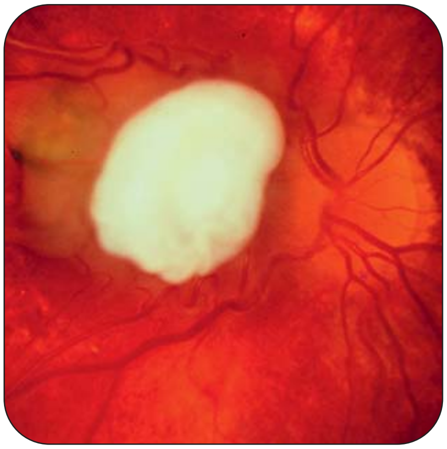

Fig. 13.262

Choroidal

Choroidal melanoma

Signs

- A typical melanoma is a pigmented, elevated, oval-shaped mass (**Fig. 13.263**).

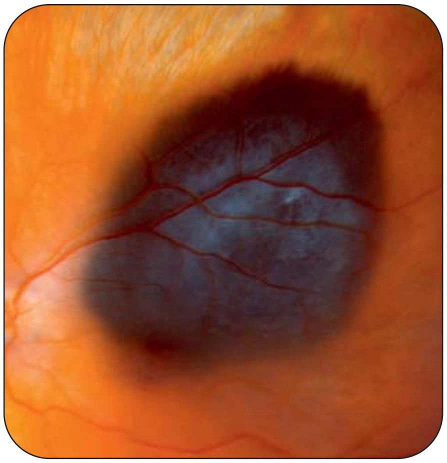

Fig. 13.263

- Often the tumour is non-pigmented (**Fig. 13.264**) and may have a mushroom-like configuration (**Fig. 13.265**).
- Orange surface lipofuscin pigment may be present (**Fig. 13.266**).

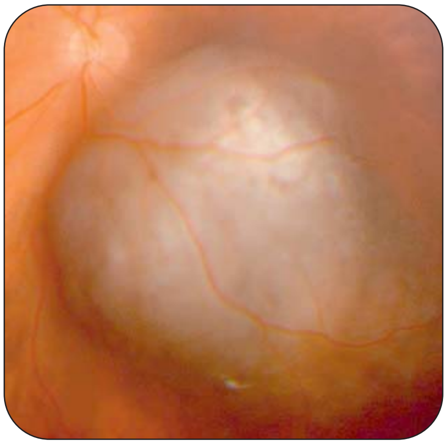
Fig. 13.264

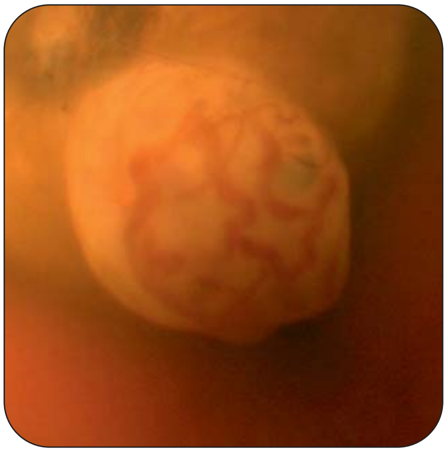
Fig. 13.265

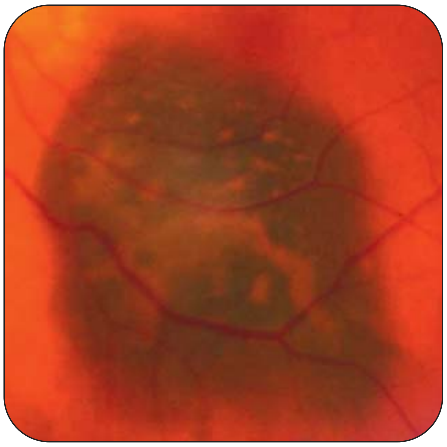
Fig. 13.266

- Ultrasonography shows acoustic hollowness within the lesion, choroidal excavation and orbital shadowing; a collar-stud configuration is pathognomonic (**Fig. 13.267**).

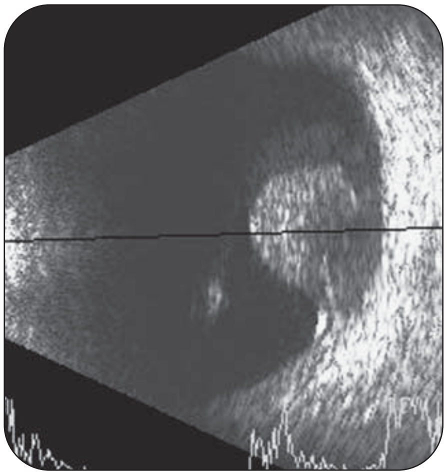
Fig. 13.267

Circumscribed choroidal haemangioma

Signs

- Oval mass in the posterior fundus with indistinct margins that has the same orange-red colour as the surrounding choroid (**Fig. 13.268**).
- Ultrasonography shows an acoustically solid lesion lacking choroidal excavation and orbital shadowing (**Fig. 13.269**).

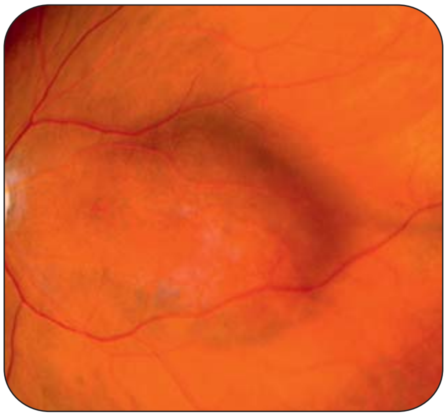
Fig. 13.268

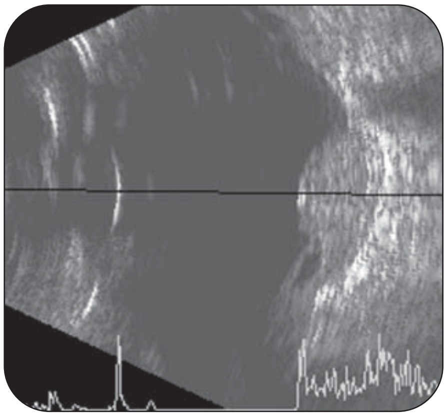
Fig. 13.269

Choroidal metastatic carcinoma

Signs

- Cream-white, placoid, oval lesion, most frequently located at the posterior pole (**Fig. 13.270**).
- Surface RPE atrophy with black clumps of pigment may be seen (**Fig. 13.271**).
- In some cases the tumour is dome-shaped and may resemble an amelanotic melanoma.
- Secondary exudative RD is common, even when the lesion is small.
- Ultrasonography of a placoid tumour shows diffuse choroidal thickening (**Fig. 13.272**).

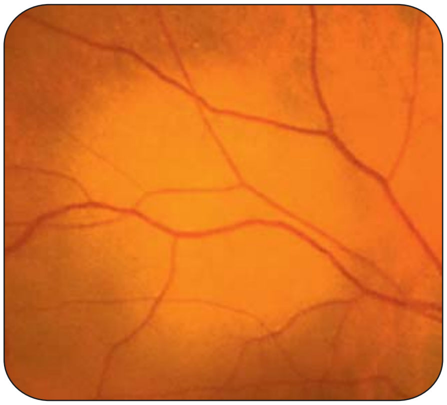
Fig. 13.270

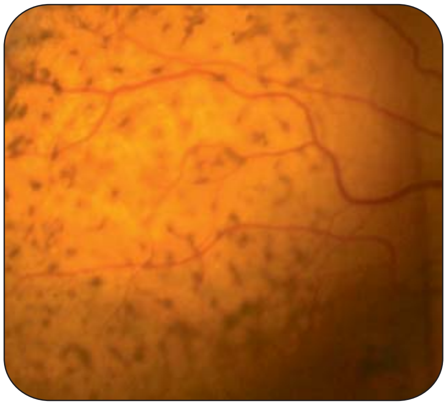
Fig. 13.271

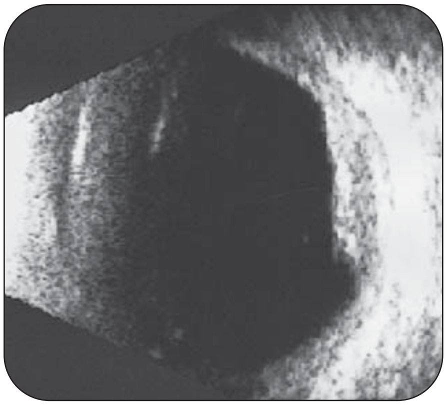
Fig. 13.272

Choroidal granuloma

Cause

- May be idiopathic or associated with sarcoidosis or tuberculosis.

Signs

- It is a dome-shaped lesion that may be associated with vitreous inflammation (**Fig. 13.273**).

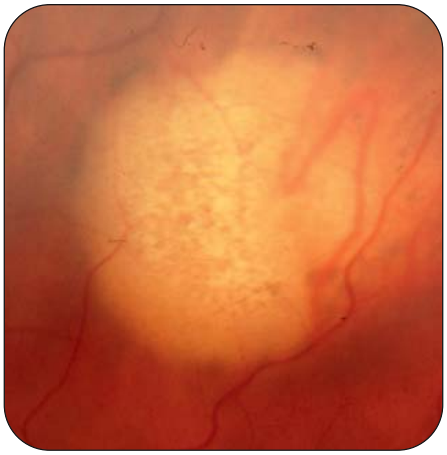
Fig. 13.273

DIFFUSE CHORIORETINAL ATROPHY

Hereditary

Choroideremia

Definition

- An XL-R disease that results in blindness by the 6th decade.

Signs

- End-stage disease shows extensive RPE and choroidal atrophy with sparing of the fovea (**Fig. 13.274**).

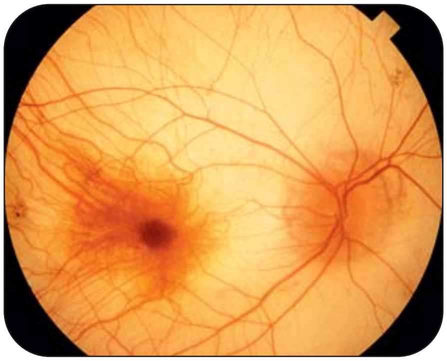

Fig. 13.274

Generalised choroidal dystrophy

Definition

- A very rare AD disease that results in blindness in adult life.

Signs

- End-stage disease is characterised by almost total choroidal atrophy with foveal involvement (**Fig. 13.275**).

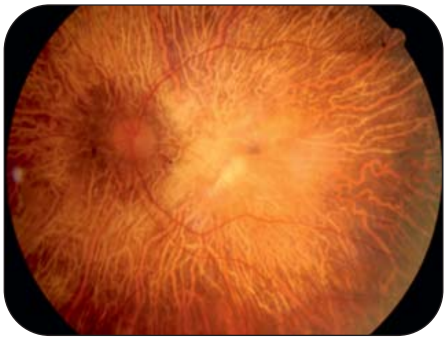

Fig. 13.275

Gyrate atrophy

Definition

- An AR disease associated with an enzyme defect that causes elevated ornithine levels.

Signs

- Mid-peripheral areas of scalloped chorioretinal atrophy (**Fig. 13.276**).
- Gradual peripheral and central progression (**Fig. 13.277**).

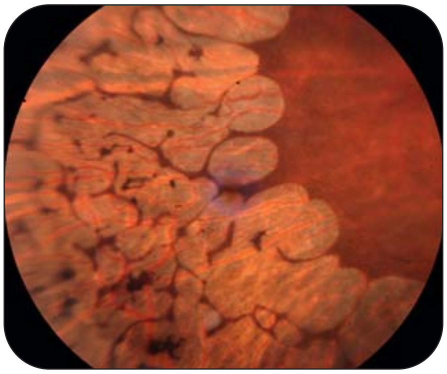

Fig. 13.276

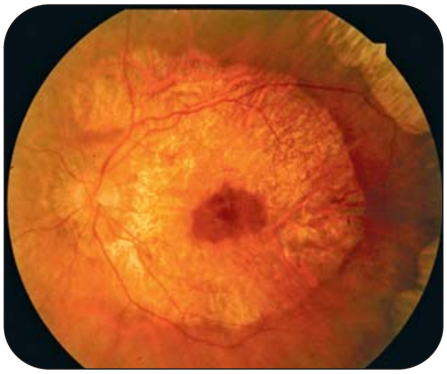

Fig. 13.277

Progressive bifocal chorioretinal atrophy

Definition

- An extremely rare AD condition that starts at birth.

Signs

- Large atrophic areas at the macula and nasally (**Fig. 13.278**).

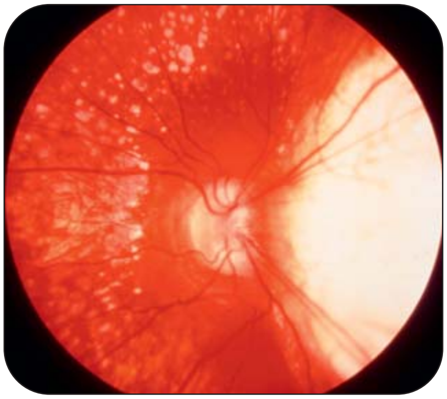

Fig. 13.278

Acquired

High myopia

Definition

- Refractive error greater than –6D and axial length greater than 26 mm.

Signs

- Large confluent areas of chorioretinal atrophy at the posterior pole are common (**Fig. 13.279**).

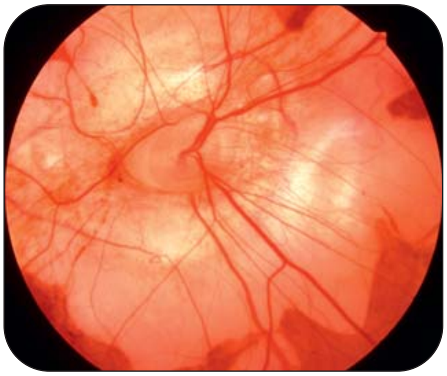

Fig. 13.279

Look for

- Lacquer cracks and CNV.
- Macular ‘coin’ haemorrhages.
- Macular hole.
- Vitreous degeneration.
- Predisposition to retinal RD, glaucoma and cataract.

Drug-induced

Chloroquine (see Fig. 13.206) and thioridazine (see Fig. 13.207).

Serpiginous chorioretinopathy

Signs

- Active disease is characterised by grey-white lesions that start around the disc and gradually spread in a serpentine manner towards the macula and peripheral fundus (see Fig. 13.112).
- Inactive end-stage disease is characterised by extensive chorioretinal atrophy (**Fig. 13.280**).

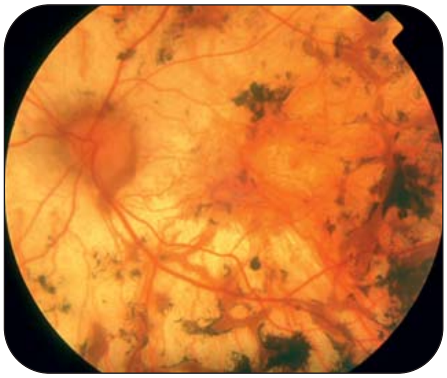

Fig. 13.280

Siderosis retinopathy

Definition

- A condition caused by dissociation of a ferrous intraocular foreign body resulting in the deposition of iron in epithelial structures.

Signs

- Retinopathy is characterised by severe atrophy of the sensory retina and RPE (**Fig. 13.281**).

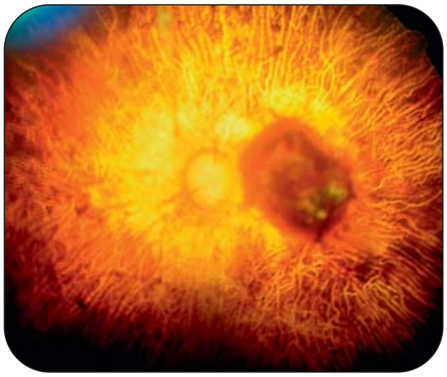

Fig. 13.281

Look for

- Cataract.
- Glaucoma.
- Iris staining that may cause heterochromia iridis (see Fig. 8.101).

Onchocercal chorioretinitis

Definition

- An infestation caused by the filarial parasite *Onchocerca volvulus* which is transmitted by the bite of a black fly.

Signs

- End-stage disease is characterised by extensive chorioretinal atrophy and arteriolar attenuation (**Fig. 13.282**).

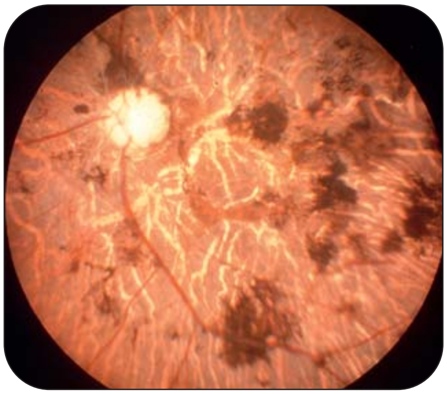

Fig. 13.282

Look for

- Anterior uveitis.
- Keratitis.

CHORIORETINAL FOLDS

Signs

- Grooves or striae at the posterior pole in which the crest is yellow and less pigmented than the darker trough.
- The folds are usually horizontal (**Fig. 13.283**) although in hypotonous eyes they radiate outwards from the disc (**Fig. 13.284**).
- FA shows alternating hyperfluorescent and hypofluorescent streaks (**Fig. 13.285**); hyperfluorescence corresponds to the crests and hypofluorescence to the troughs.

Causes

a. *Idiopathic folds* affect both eyes of healthy hypermetropic patients.

b. *Orbital disease* – retrobulbar tumours and thyroid ophthalmopathy.

c. *Miscellaneous* – choroidal tumours, chronic papilloedema, posterior scleritis, ocular hypotony and scleral buckling for RD.

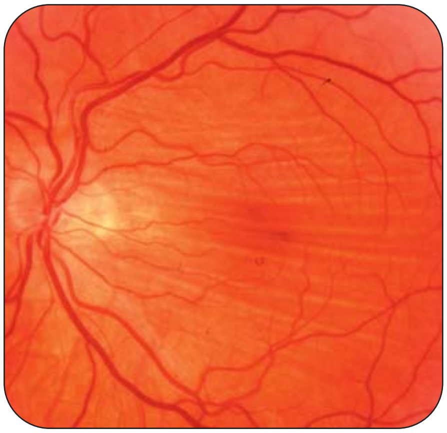

Fig. 13.283

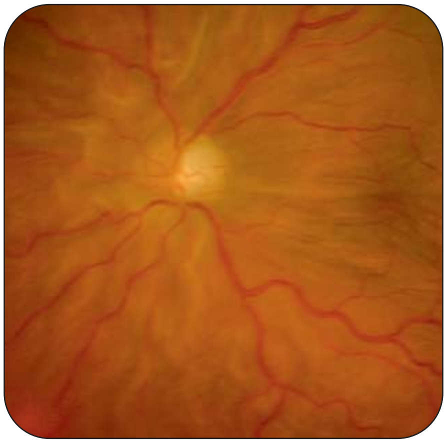

Fig. 13.284

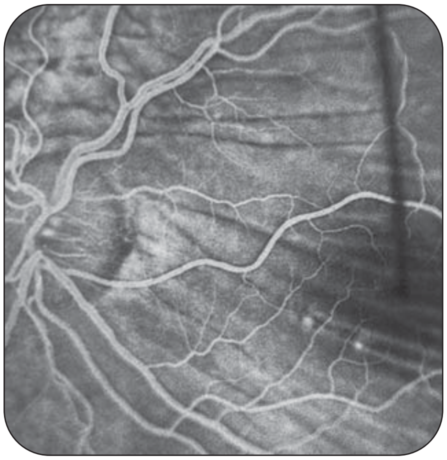

Fig. 13.285

DIFFERENTIAL DIAGNOSIS OF VISUAL SYMPTOMS

Table 13.7 **Poor vision but normal fundus**

1. **In adults**
   - Retrobulbar neuritis – check pupillary reactions and colour vision
   - Nutritional amblyopia – check colour vision
   - Cancer-associated retinopathy – fundus may be initially normal
2. **In children**
   - Refractive error
   - Amblyopia – check refraction and look for squint
   - Non-organic visual loss
   - Delayed visual maturation – vision improves to normal by 6 months
   - Early retinal dystrophies – cone and Stargardt
   - Cortical visual impairment
   - Leber congenital amaurosis – roving eye movements are poorly reacting pupils to light
   - Achromatopsia (rod monochromatism) – nystagmus and photophobia
   - X-linked recessive congenital stationary night-blindness – nystagmus, nyctalopia and myopia

Table 13.8 **Transient monocular visual loss**

1. **Retinal vascular**
   - Embolism
   - Vasospasm (retinal migraine)
   - Hypoperfusion due to carotid stenosis
   - Impending CRV or CRA occlusion
2. **Optic nerve**
   - Impending AION
   - Papilloedema
   - Optic disc drusen
3. **Ocular**
   - Intermittent angle-closure glaucoma
   - Recurrent hyphaema

Ocular motility 14

CONCOMITANT ESOTROPIA 357

Non-accommodative 357
- *Early-onset (infantile)* 357
- *Basic near* 357
- *Distance* 358
- *Microtropia* 358
- *Acute (late onset)* 358
- *Sensory (secondary)* 358
- *Convergence spasm* 359
- *Cyclic* 359
- *Consecutive* 359

Refractive accommodative 360
- *Fully accommodative* 360
- *Partly accommodative* 360

Non-refractive accommodative 360
- *Convergence excess* 360
- *Hypoaccommodative convergence excess* 361

Pseudo-esotropia 361
- *Epicanthic folds and broad nasal bridge* 361
- *Negative angle kappa* 361
- *Narrow interpupillary distance* 361

CONCOMITANT EXOTROPIA 361
- *Constant (early onset)* 361
- *Intermittent (basic)* 362
- *Sensory (secondary)* 362
- *Consecutive* 362
- *Microexotropia* 362
- *Hypertelorism* 362
- *Pseudo-exotropia* 363

CONGENITAL SYNDROMES 363
- *Duane retraction syndrome type 1* 363
- *Duane retraction syndrome type 2* 363
- *Duane retraction syndrome type 3* 364
- *Brown syndrome* 364
- *Monocular elevator deficit* 365
- *Möbius syndrome* 365
- *Congenital fibrosis syndrome* 365
- *Strabismus fixus* 366

ALPHABET PATTERNS 366
- *'V' pattern* 366
- *'A' pattern* 366
- *'X' pattern* 366
- *'Y' pattern* 367
- *'Inverted Y' (lambda) pattern* 367

NEUROGENIC PALSIES 367

Oculomotor nerves 367
- *Third nerve palsy* 367
- *Fourth nerve palsy* 368
- *Sixth nerve palsy* 369

Individual muscle palsies 370
- *Inferior rectus palsy* 370
- *Superior rectus palsy* 370
- *Inferior oblique palsy* 371

TORSIONAL DEVIATIONS IN PRIMARY POSITION 371
- *Incyclotorsion* 371
- *Excyclotorsion* 371

MUSCLE OVERACTIONS 372

Up-shoot in adduction 372
- *Primary inferior oblique overaction* 372
- *Secondary inferior oblique overaction* 372

Up-shoot in abduction 372

Down-shoot in adduction 372

Down-shoot in abduction 373

ACQUIRED MYOGENIC AND RESTRICTIVE PALSIES 373
- *Restrictive thyroid myopathy* 373
- *Myasthenia gravis* 373
- *Medial wall blow-out fracture* 374
- *Orbital floor blow-out fracture* 374
- *Acute myositis* 374
- *Frozen orbit* 374
- *Chronic progressive external ophthalmoplegia* 375
- *Iatrogenic palsies* 375

GAZE PALSIES 376

Horizontal gaze palsy 376
- *Medial longitudinal fasciculus lesion* 376
- *Paramedian pontine reticular formation lesion* 376
- *Combined MLF and ipsilateral PPRF (or abducens nucleus) lesion* 376
- *Bilateral pontine lesions* 376

Vertical gaze palsy 377
- *Cerebrovascular accidents* 377
- *Parinaud dorsal midbrain syndrome* 377
- *Progressive supranuclear palsy (Steele–Richardson–Olszewski syndrome)* 377

NYSTAGMUS 378
- *Physiological* 378
- *Primary congenital* 378
- *Acquired ocular* 379
- *Peripheral vestibular* 380
- *Sidebeat* 380
- *Spasmus nutans* 381
- *Upbeat* 381
- *Downbeat* 382
- *Convergence-retraction* 382

CONCOMITANT ESOTROPIA

In a comitant deviation the angle is the same, or nearly the same, for all positions of gaze; the two types are non-accommodative and accommodative.

Non-accommodative

Early-onset (infantile)

Presentation

- Before the age of 6 months.

Signs

- Large and stable angle with no significant refractive error.
- Fixation may be alternating in the primary position with cross fixation in side-gaze (**Figs 14.1** and **14.2**).

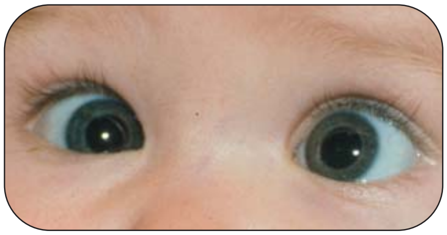

Fig. 14.1

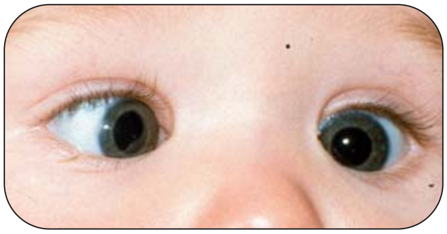

Fig. 14.2

Look for

- Latent and manifest-latent nystagmus.
- Inferior oblique overaction may be present initially or develop later.
- Dissociated vertical deviation (DVD) which often develops by the age of 3 years.

Differential diagnosis (Table 14.1)

Table 14.1 **Differential diagnosis of early-onset esotropia**

- Congenital bilateral sixth nerve palsy
- Secondary (sensory) esotropia (see Figs 14.3 and 14.5)
- Nystagmus blockage syndrome in which convergence dampens a horizontal nystagmus
- Duane syndrome types I and III (see Figs 14.16 and 14.17)
- Möbius syndrome
- Strabismus fixus

Basic near

Presentation

- After the age of 6 months.

Signs

- Straight for distance and esotropia for near.
- Normal or low AC/A ratio and normal refraction.

Distance

Presentation

- Early adult life.

Signs

- Intermittent or constant esotropia for distance, and minimal or no deviation for near.
- Myopia is common.

Microtropia

Presentation

- May be spontaneous or following surgery for a large deviation.

Signs

- Very small angle that may not be apparent on cover testing.
- Central suppression scotoma of the deviating eye.
- Abnormal retinal correspondence (ARC) with reduced stereopsis and variable peripheral fusional amplitudes.
- Anisometropia and amblyopia are common.

Acute (late onset)

Presentation

- At about 5 years of age with sudden onset of diplopia and esotropia.

Signs

- Esotropia for distance and near.
- Normal motility.
- Normal refraction.

Differential diagnosis

- Sixth nerve palsy.

Sensory (secondary)

Pathogenesis

- Unilateral congenital or early-onset reduction in visual acuity which interferes with or abolishes fusion.

Look for

- Opacity of the media such as cataract.
- Optic atrophy or optic nerve hypoplasia.
- Colobomatous microphthalmos (**Fig. 14.3**).
- Macular scarring from toxoplasmosis (**Fig. 14.4**).
- Retinoblastoma (**Fig. 14.5**).

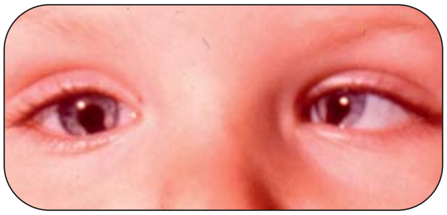
Fig. 14.3

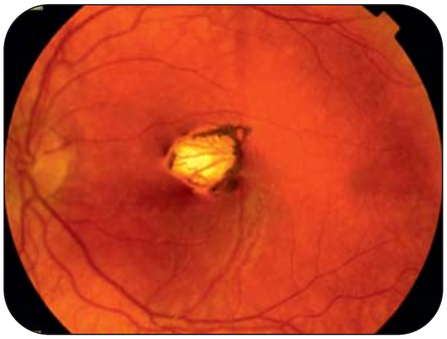
Fig. 14.4

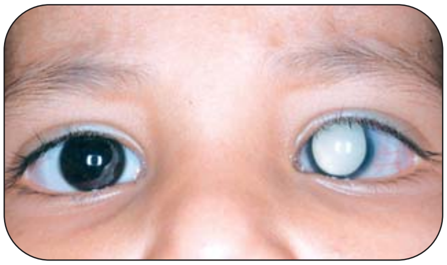
Fig. 14.5

Convergence spasm

Presentation

- Usually in adult life as an intermittent phenomenon which is usually hysterical.

Signs

- Esotropia and bilateral miosis (**Fig. 14.6**).
- Pseudo-myopia due to accommodative spasm.

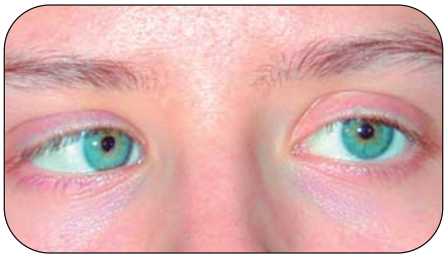
Fig. 14.6

Cyclic

Presentation

- Alternating manifest esotropia lasting 24 hours followed by normal binocularity for 24 hours.

Signs

- Large deviation but diplopia is unusual.
- Fusional amplitudes are subnormal or absent.

Consecutive

Cause

- Surgical overcorrection of exotropia.

Refractive accommodative

The AC/A ratio is normal and esotropia is a physiological response to excessive hypermetropia. Presentation is usually between 2 and 3 years of age.

Fully accommodative

Signs

- Esotropia without correction (**Fig. 14.7A**).
- Straight after correction of hypermetropia (**Fig. 14.7B**).

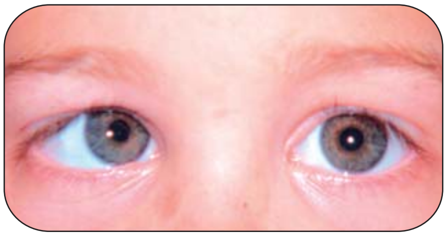
Fig. 14.7A

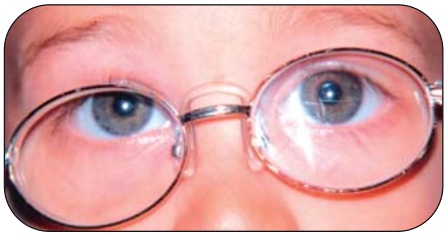
Fig. 14.7B

Partly accommodative

Signs

- Esotropia without correction (**Fig. 14.8A**).
- Angle is reduced but not eliminated after correction of hypermetropia (**Fig. 14.8B**).

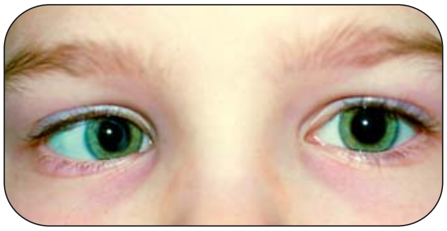
Fig. 14.8A

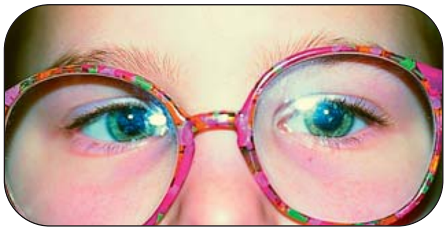
Fig. 14.8B

Non-refractive accommodative

Refraction is normal but the AC/A ratio is high so that a unit increase of accommodation is accompanied by a disproportionately large increase in convergence.

Convergence excess

AC/A ratio is high due to increased accommodative convergence.

Signs

- Straight for distance (**Fig. 14.9A**).
- Esotropia for near (**Fig. 14.9B**).
- Straight through bifocals (**Fig. 14.9C**).

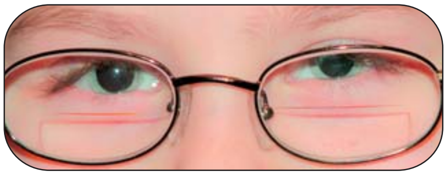
Fig. 14.9A

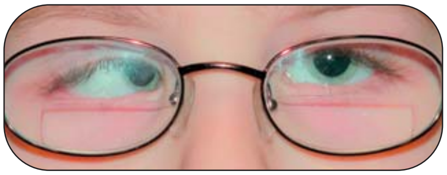
Fig. 14.9B

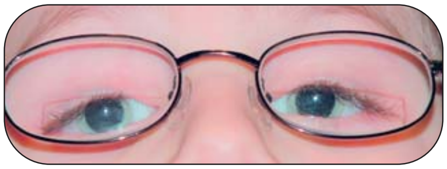
Fig. 14.9C

Hypoaccommodative convergence excess

AC/A ratio is high due to weak accommodation.

Signs

- Remote near point of accommodation.
- Straight for distance.
- Esotropia for near.

Pseudo-esotropia

Epicanthic folds and broad nasal bridge

This may simulate or accentuate an esotropia.

Signs

- Symmetric corneal light reflexes are central or slightly displaced nasally (**Fig. 14.10**).
- No deviation on cover-uncover test.

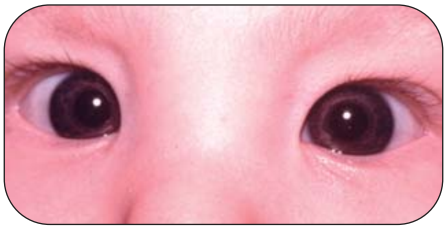

Fig. 14.10

Negative angle kappa

Signs

- The corneal light reflexes are displaced temporally as may occur in high myopia.

Narrow interpupillary distance

This may occur in hypotelorism in which there is narrow separation of the orbits as a result of skull malformation or failure of brain development.

CONCOMITANT EXOTROPIA

Constant (early onset)

Presentation

- Often at birth.

Signs

- Normal refraction.
- Large and constant angle.
- DVD may be present.

Look for

- Neurological anomalies.

Intermittent (basic)

Presentation

- Around 2 years of age with exophoria breaking down to exotropia.

Signs

- Straight with BSV at times (**Fig. 14.11A**).
- Divergent with suppression at other times (**Fig. 14.11B**).

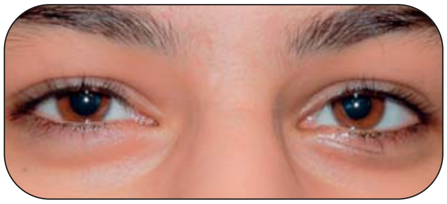

Fig. 14.11A

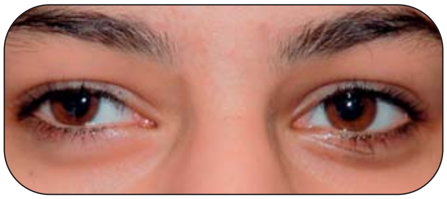

Fig. 14.11B

Classification

a. *Distance* (divergence excess) – angle is greater for distance.

b. *Near* (convergence insufficiency) – angle is greater for near.

c. *Non-specific* – angle is the same for distance and near.

Sensory (secondary)

Pathogenesis

- Monocular visual impairment by acquired lesions such as cataract (**Fig. 14.12**) or other opacities of the media.

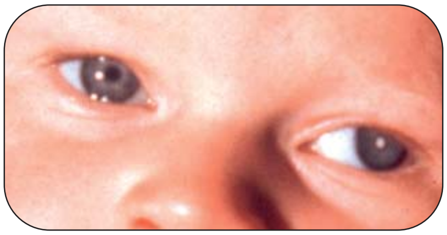

Fig. 14.12

Consecutive

Presentation

- In adult life either in an amblyopic eye or following surgery for esotropia.

Signs

- Large and constant deviation (**Fig. 14.13**).

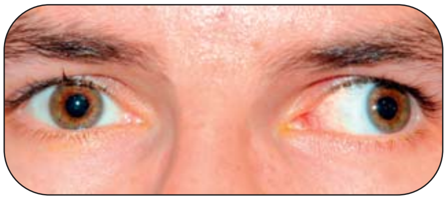

Fig. 14.13

Microexotropia

Presentation

- In isolation or in association with a primary intermittent or constant exotropia.

Look for

- Amblyopia and anisometropia.

Hypertelorism

Signs

- Congenital anomaly in which there is wide separation of the orbits often associated with shallow orbits as in Crouzon syndrome.
- Gross exotropia (**Fig. 14.14**).

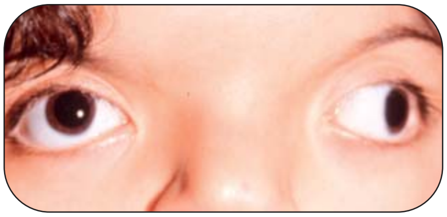

Fig. 14.14

Pseudo-exotropia

a. *Positive angle kappa without ocular anomalies* in which the corneal light reflexes are displaced nasally giving a false impression of an exotropia.

b. *Positive angle kappa* is caused by macular heterotopia (e.g. dragged macula in retinopathy of prematurity – **Fig. 14.15**) with temporal displacement of the fovea and consequent nasal displacement of the corneal light reflexes.

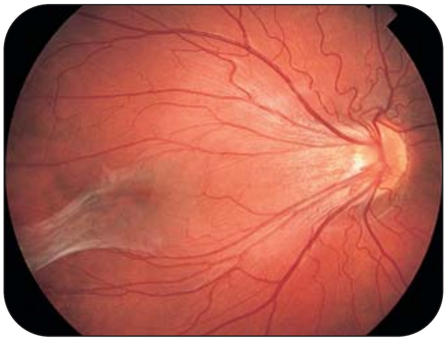

Fig. 14.15

CONGENITAL SYNDROMES

Duane retraction syndrome type 1

Signs (left eye)

- Primary position – straight or slight esotropia (**Fig. 14.16A**).
- Abduction is absent or limited and is accompanied by widening of the palpebral fissure (**Fig. 14.16B**).
- Adduction is normal or mildly limited and is accompanied by retraction of the globe and narrowing of the palpebral fissure (**Fig. 14.16C**).

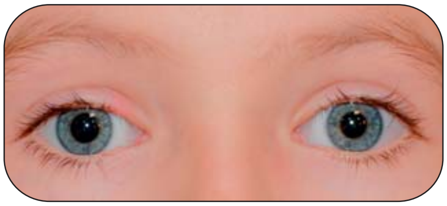

Fig. 14.16A

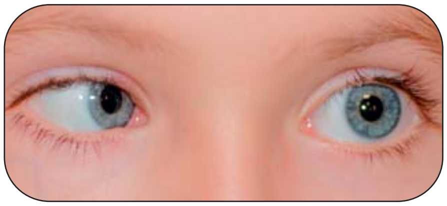

Fig. 14.16B

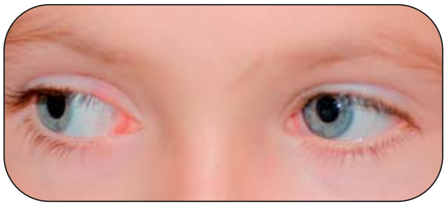

Fig. 14.16C

Duane retraction syndrome type 2

Signs

- Primary position – straight or slight exotropia.
- Abduction is normal or slightly limited.
- Adduction is limited.

Duane retraction syndrome type 3

Signs (left eye)

- Primary position – straight or slight esotropia (**Fig. 14.17A**).
- Abduction (**Fig. 14.17B**) and adduction (**Fig. 14.17C**) are both limited.
- All three types of Duane syndrome may be associated with up-shoot of the affected eye on adduction and occasionally a down-shoot.

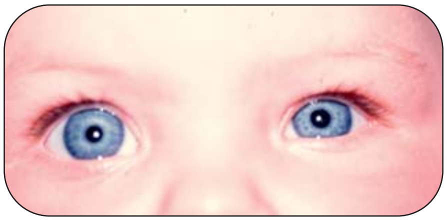
Fig. 14.17A

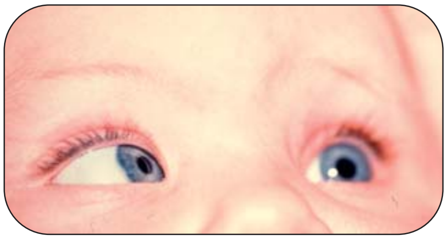
Fig. 14.17B

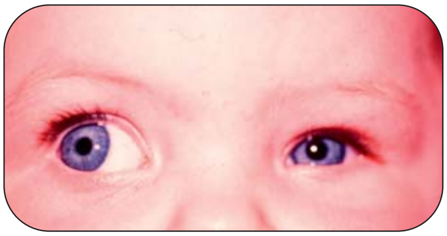
Fig. 14.17C

Brown syndrome

Signs (left eye)

- Primary position – straight (**Fig. 14.18A**).
- Limited elevation in adduction (**Fig. 14.18B**).
- Limited elevation on upgaze (**Fig. 14.18C**).
- Normal left elevation in abduction (**Fig. 14.18D**).
- Absence of left superior oblique overaction.

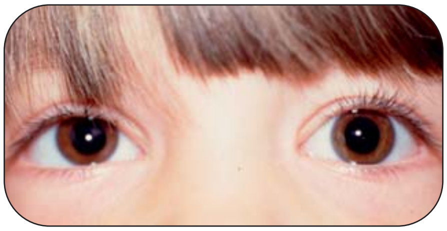
Fig. 14.18A

Causes (Table 14.2)

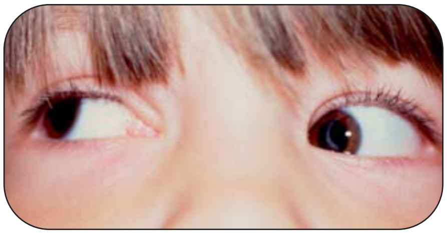
Fig. 14.18B

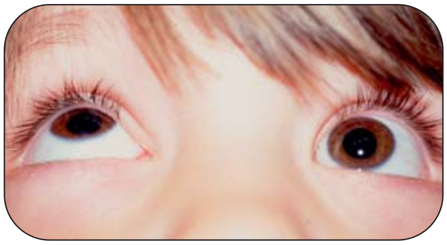
Fig. 14.18C

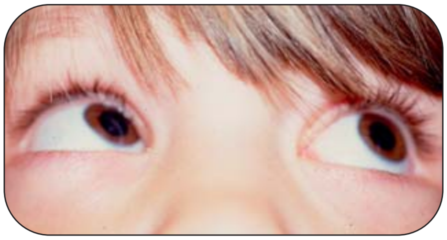
Fig. 14.18D

Table 14.2 **Causes of Brown syndrome**

1. **Congenital**
   - Idiopathic
   - Congenital click syndrome – impaired movement of the superior oblique tendon through the trochlea
2. **Acquired**
   - Trauma to the trochlea or superior oblique tendon
   - Tendonitis associated with rheumatoid arthritis, pansinusitis and scleritis

Monocular elevator deficit

Signs (right eye)

- Primary position – usually straight.
- Profound limited elevation in all positions of upgaze (**Fig. 14.19A–C**).
- Normal abduction and adduction.
- Normal depression in all positions.

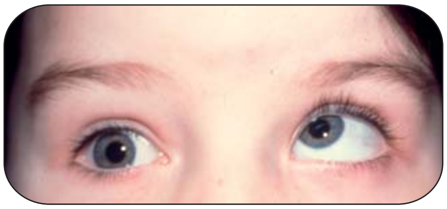

Fig. 14.19A

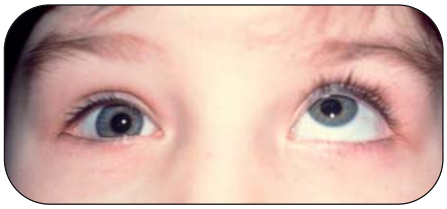

Fig. 14.19B

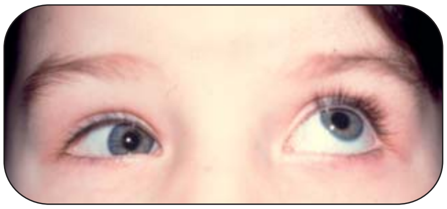

Fig. 14.19C

Möbius syndrome

Signs

- Primary position – usually straight or esotropia.
- Unilateral or bilateral abduction deficit or horizontal gaze palsy.

Look for

- Bilateral facial paresis with the mouth constantly held open (**Fig. 14.20**).
- Paresis of the 9th and 12th cranial nerves; the latter results in atrophy of the tongue (**Fig. 14.21**).
- Mild mental handicap.
- Limb anomalies.
- Deafness.

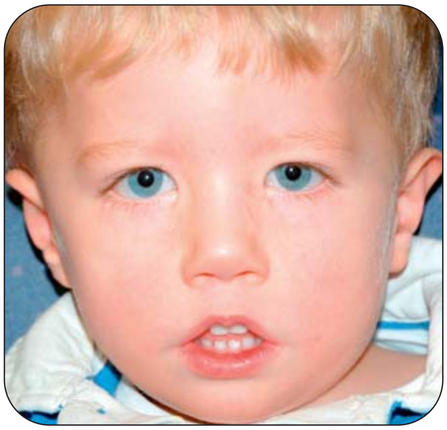

Fig. 14.20

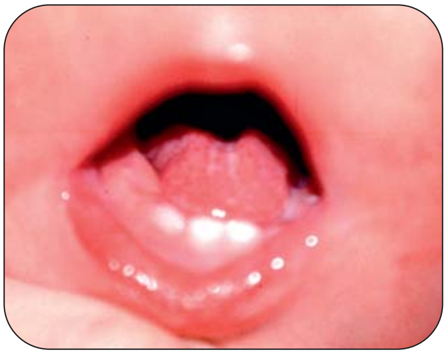

Fig. 14.21

Congenital fibrosis syndrome

Signs

- Severe bilateral ptosis (**Fig. 14.22**).
- In the primary position the eyes are fixed below the horizontal by about 10°.
- The eyes are usually divergent but may be convergent or straight.
- Severe restriction of vertical gaze and variable restriction of horizontal gaze.
- Compensatory head posture.

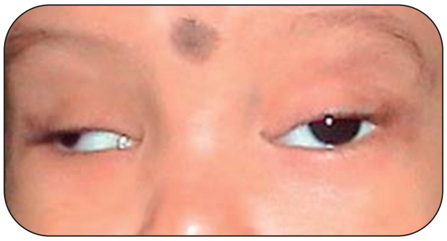

Fig. 14.22

Strabismus fixus

Signs

- Both eyes are fixed by fibrous tightening of the medial recti (convergent strabismus fixus – **Fig. 14.23**), or the lateral recti (divergent strabismus fixus – **Fig. 14.24**).

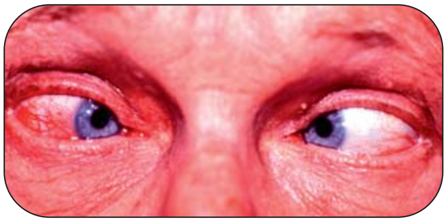
Fig. 14.23

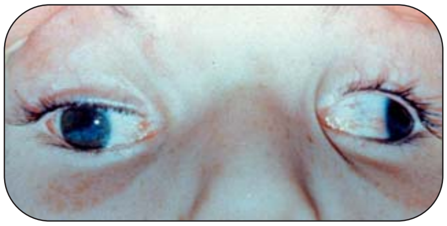
Fig. 14.24

ALPHABET PATTERNS

'V 'or 'A' patterns may occur when the relative contributions of the superior rectus and inferior oblique to elevation, or of the inferior rectus and superior oblique to depression are abnormal. This results in abnormal balance of their horizontal vectors in up- and downgaze.

'V' pattern

Signs

- Difference between upgaze and downgaze is ≥15Δ.
- Primary position – esotropia (**Fig. 14.25**) or exotropia (**Fig. 14.26**).

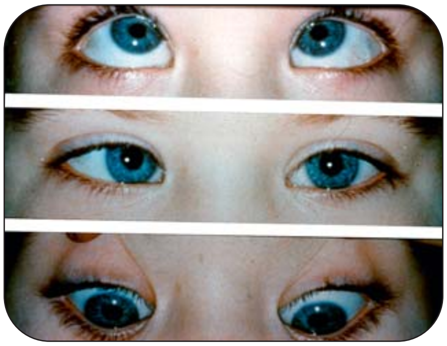
Fig. 14.25

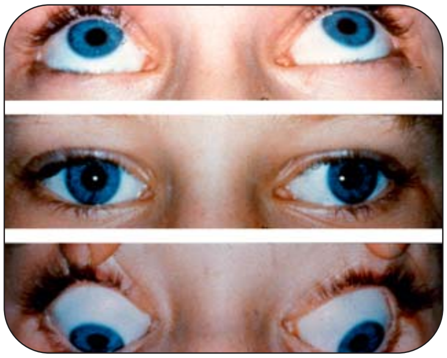
Fig. 14.26

'A' pattern

Signs

- Difference between upgaze and downgaze is ≥10Δ.
- Primary position – esotropia (**Fig. 14.27**) or exotropia (**Fig. 14.28**).

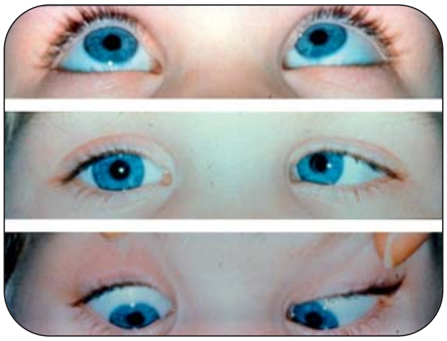
Fig. 14.27

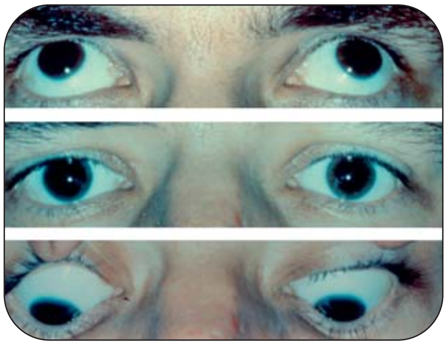
Fig. 14.28

'X' pattern

Signs

- Orthophoria in the primary position but exotropia in upgaze and downgaze usually associated with both inferior and superior oblique overaction.

‘Y’ pattern

Signs

- Divergence only in upgaze.

Inverted ‘Y’ (lambda) pattern

Signs

- Divergence only in downgaze.

NEUROGENIC PALSIES

Oculomotor nerves

Third nerve palsy

Signs (right eye)

- Profound ptosis (**Fig. 14.29A**).
- Abducted eye in the primary position (**Fig. 14.29B**).
- Normal abduction (**Fig. 14.29C**).
- Limited adduction (**Fig. 14.29D**).
- Limited depression (**Fig. 14.29E**) and elevation (**Fig. 14.29F**).
- Intact superior oblique muscle causes intorsion of the eye at rest which increases on attempted downgaze.
- Parasympathetic involvement causing mydriasis and cycloplegia.

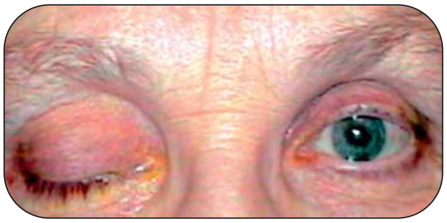
Fig. 14.29A

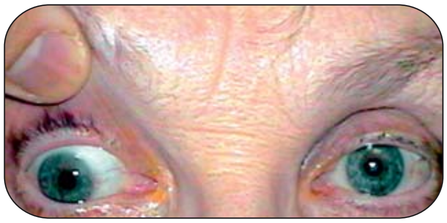
Fig. 14.29B

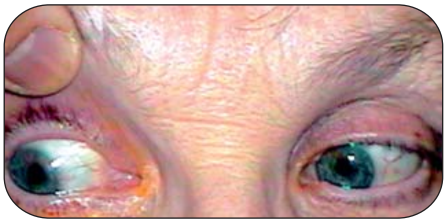
Fig. 14.29C

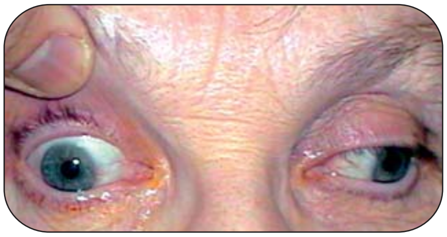
Fig. 14.29D

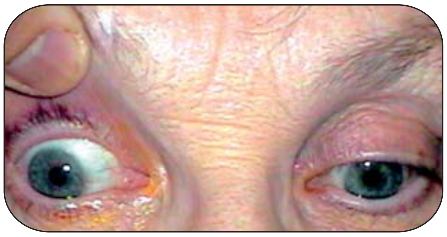
Fig. 14.29E

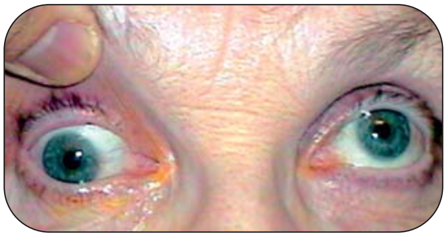
Fig. 14.29F

Causes of isolated third nerve palsy

a. *Idiopathic* – about 25%.

b. *Vascular* – hypertension and diabetes.

c. *Aneurysm* of the posterior communicating artery at its junction with the internal carotid artery.

d. *Trauma* – both direct and secondary to subdural haematoma with uncal herniation.

e. *Miscellaneous* – tumours, syphilis, giant cell arteritis and other collagen vascular disorders.

Fourth nerve palsy

Signs (left eye)

- Primary position – left hypertropia ('left over right') with the uninvolved right eye fixating (**Fig. 14.30A**).
- Increase of left hypertropia on right gaze due to inferior oblique overaction (**Fig. 14.30B**).
- Limitation of left depression in adduction (**Fig. 14.30C**).
- Normal – left abduction (**Fig. 14.30D**), depression (**Fig. 14.30E**) and elevation (**Fig. 14.30F**).
- Excyclotorsion.
- Diplopia – vertical, torsional and worse on downgaze.

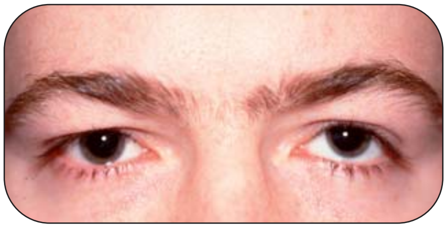
Fig. 14.30A

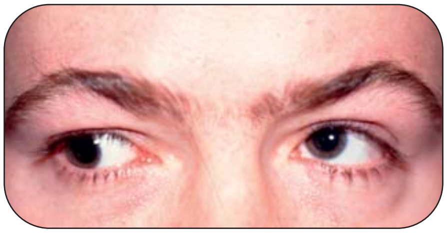
Fig. 14.30B

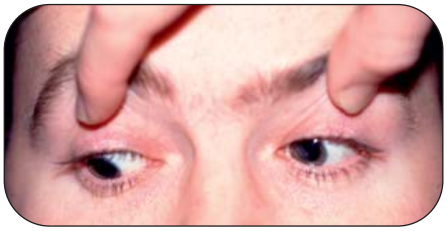
Fig. 14.30C

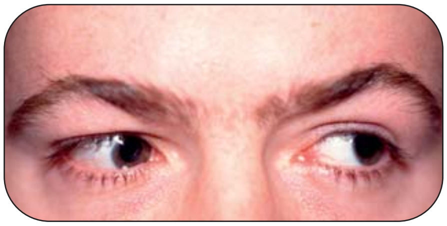
Fig. 14.30D

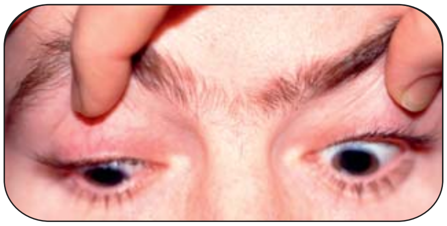
Fig. 14.30E

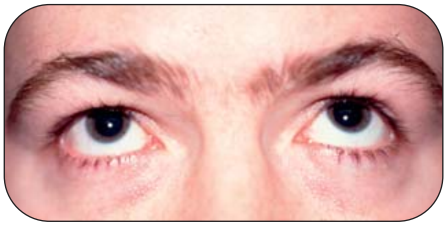
Fig. 14.30F

Head posture (Fig. 14.31)

- Head tilt – to right.
- Face turn – to left.
- Chin – slightly depressed.

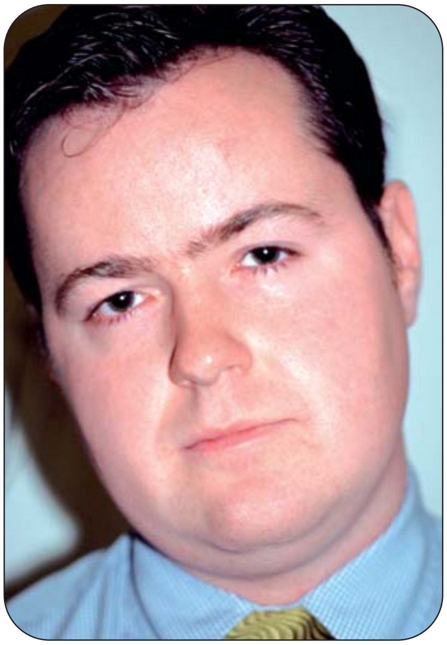

Fig. 14.31

Parks three step test

a. First step – which eye is hypertropic in the primary position?

- Left hypertropia may be caused by weakness of one of the following four muscles: one of the depressors of the left eye (superior oblique or inferior rectus) or one of the elevators of the right eye (superior rectus or inferior oblique).
- In 4th nerve palsy the involved eye is higher.

b. Step two – is left hypertropia greater in right gaze or left gaze?

- Increase on right gaze implicates either the right superior rectus or left superior oblique.
- Increase on left gaze implicates either the right inferior oblique or left inferior rectus. (In 4th nerve palsy the deviation is **W**orse **O**n **O**pposite **G**aze – **WOOG**.)

c. Step three – Bielschowsky head tilt test

- The head is tilted to the right and then to the left.
- Increase of left hypertropia on left head tilt (**Fig. 14.32A**) and decrease on right head tilt (**Fig. 14.32B**) implicates the left superior oblique.
- Increase of right hypertropia on left head tilt implicates the right inferior rectus. (In 4th nerve palsy the deviation is **B**etter **O**n **O**pposite **T**ilt – **BOOT**.)

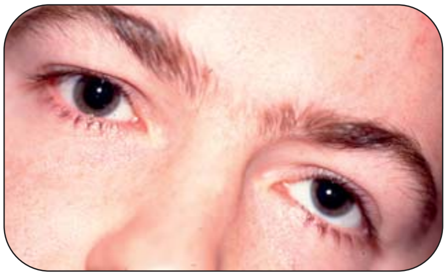

Fig. 14.32A

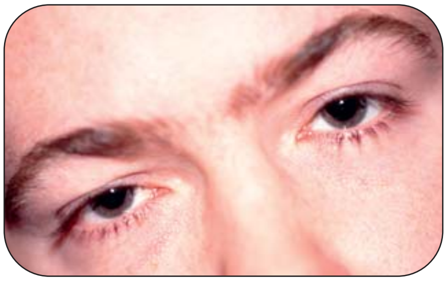

Fig. 14.32B

Causes of isolated fourth nerve palsy

a. *Congenital* (common).

b. *Closed head trauma* – often bilateral palsy.

c. *Vascular*.

d. *Intracranial tumour* (rare).

e. *Intracranial aneurysm* (rare).

f. *Iatrogenic* – displacement of the trochlea during orbital surgery.

Sixth nerve palsy

Signs (right eye)

a. Acute

- Primary position – right esotropia (**Fig. 14.33A**).
- Marked limitation of right abduction (**Fig. 14.33B**).
- Forced duction test – negative.

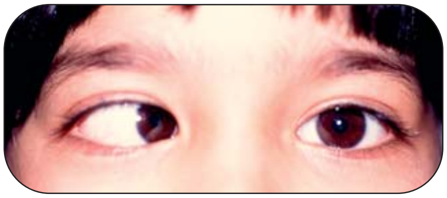

Fig. 14.33A

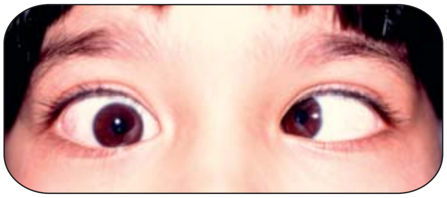

Fig. 14.33B

b. Long-standing

- Primary position – straight or slight esotropia (**Fig. 14.34A**).
- Limitation of right abduction (**Fig. 14.34B**).
- Forced duction test – positive due to contracture of the ipsilateral medial rectus.

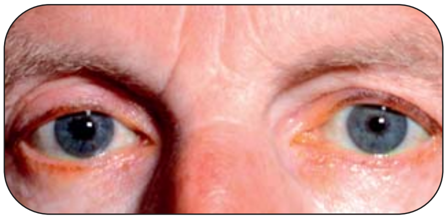

Fig. 14.34A

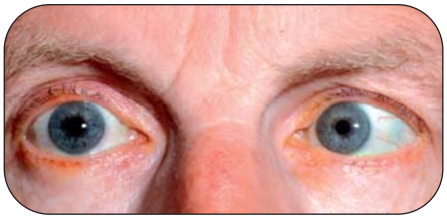

Fig. 14.34B

Individual muscle palsies

Inferior rectus palsy

Signs (left eye)

- Primary position – left hypertropia.
- Left underaction on depression in abduction and right down-shoot due to superior oblique overaction (**Fig. 14.35**).
- Incyclotorsion.

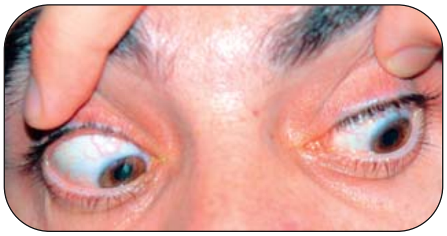

Fig. 14.35

Head posture

- Head tilt – to right.
- Face turn – to left.
- Chin – depressed.

Bielschowsky test

- Increase in left hypertropia on right head tilt.

Superior rectus palsy

Signs (left eye)

- Primary position – left hypotropia.
- Left underaction on elevation in abduction and right up-shoot due to inferior oblique overaction (**Fig. 14.36**).
- Small excyclotorsion.

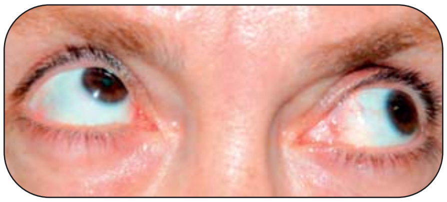

Fig. 14.36

Head posture

- Head tilt – usually contralateral but may be to either side.
- Face turn – to left if recent onset.
- Chin – elevation if recent onset.

Bielschowsky test

- Increase in left hypotropia on left head tilt.

Inferior oblique palsy

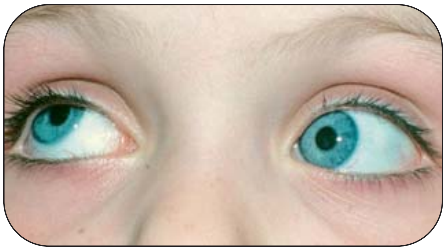

Fig. 14.37

Signs (left eye)

- Primary position – left hypotropia.
- Left underaction on elevation in adduction and right up-shoot due to superior rectus overaction (**Fig. 14.37**).
- Incyclotorsion.

Head posture

- Head tilt – to left.
- Face turn – to right.
- Chin – elevated.

Bielschowsky test

- Increase in left hypotropia on right head tilt.

TORSIONAL DEVIATIONS IN PRIMARY POSITION

Incyclotorsion

Causes

- Inferior rectus palsy.
- Inferior oblique palsy.

Excyclotorsion

Causes

- Superior oblique palsy.
- Superior rectus palsy.
- Primary inferior oblique overaction.
- Skew deviation in which the hypotropic eye is excyclotorted.

MUSCLE OVERACTIONS

Up-shoot in adduction

Primary inferior oblique overaction

Signs

- Frequently bilateral (**Figs 14.38A** and **B**).
- 'V' esotropia (**Figs 14.38C** and **D**).
- Negative Bielschowsky test.

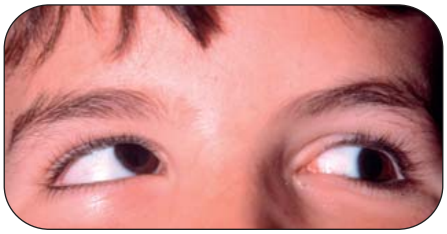

Fig. 14.38A

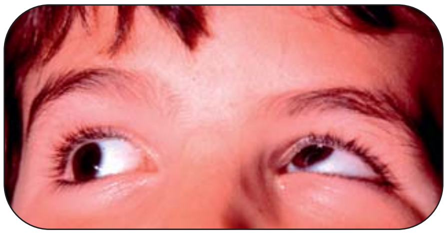

Fig. 14.38B

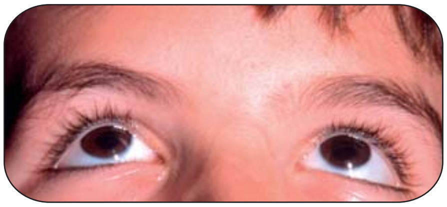

Fig. 14.38C

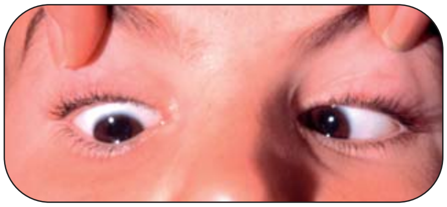

Fig. 14.38D

Secondary inferior oblique overaction

Causes

- With ipsilateral superior oblique palsy (see Fig. 14.30B).
- With contralateral superior rectus palsy (see Fig. 14.36).

Up-shoot in abduction

Causes

- Inferior rectus palsy with ipsilateral superior rectus overaction.
- Inferior oblique palsy with contralateral superior rectus overaction (see Fig. 14.37).

Down-shoot in adduction

Causes

- Duane syndrome.
- Brown syndrome.
- Secondary superior oblique overaction due to ipsilateral inferior oblique palsy or contralateral inferior rectus palsy (see Fig. 14.35).

Down-shoot in abduction

Causes

- Excyclorotation of the orbit and contents is seen in certain craniofacial syndromes (**Fig. 14.39A**).
- If the globe is excyclorotated the lateral rectus acts not only as an abductor but also as a depressor (**Fig. 14.39B** and **C**).

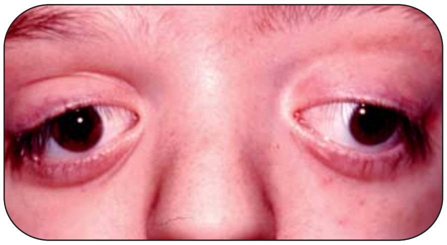

Fig. 14.39A

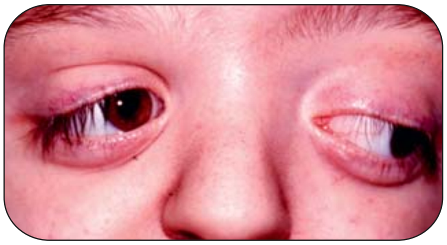

Fig. 14.39B

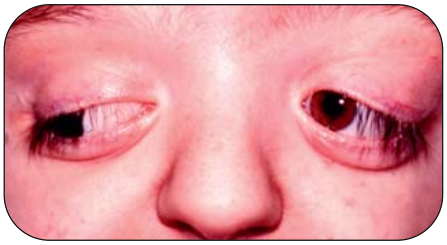

Fig. 14.39C

ACQUIRED MYOGENIC AND RESTRICTIVE PALSIES

Restrictive thyroid myopathy

Restrictive myopathy causes limitation in the direction opposite to the field of action of the involved muscle.

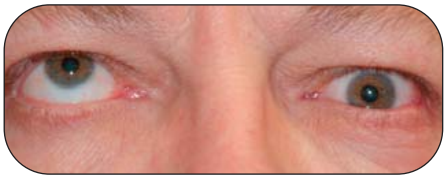

Fig. 14.40A

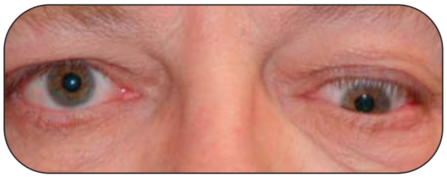

Fig. 14.40B

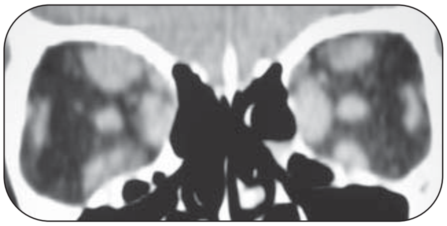

Fig. 14.41

Signs

- The most frequently involved muscle is the inferior rectus (causing limited elevation – **Fig. 14.40A**).
- Next is the medial rectus (limiting abduction), followed by the superior rectus (limiting depression) and the lateral rectus (limiting adduction).

Look for

- Lid retraction and lag (**Fig. 14.40B**).
- Chemosis and periocular oedema.
- Superior limbic keratoconjunctivitis.
- Proptosis.
- CT showing muscle enlargement (**Fig. 14.41**).

Myasthenia gravis

Myasthenia may involve any or all of the extraocular muscles and may even mimic internuclear ophthalmoplegia but the most frequent motility defect is limitation of elevation.

Look for

- Ptosis (**Fig. 14.42**).
- Excessive muscular fatigue.
- Lack of facial expression.
- Dysphagia and dysarthria.

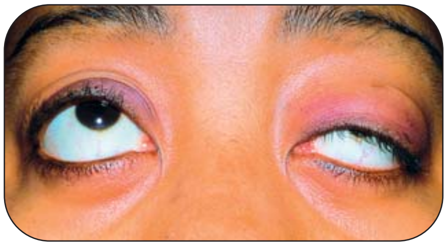

Fig. 14.42

Medial wall blow-out fracture

Signs

- Entrapment within the fracture of the medial rectus muscle causes limited abduction (**Fig. 14.43A**) and occasionally adduction (**Fig. 14.43B**).

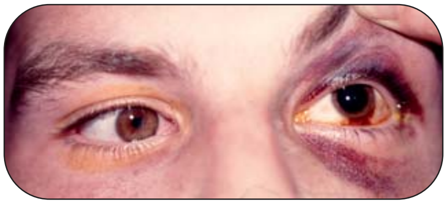

Fig. 14.43A

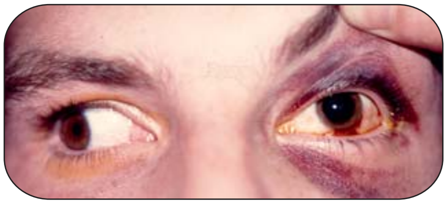

Fig. 14.43B

Look for

- Subcutaneous emphysema (see Fig. 1.39).

Orbital floor blow-out fracture

Signs

- Entrapment of the inferior rectus and inferior oblique give rise to limited elevation (**Fig. 14.44A**).
- CT is useful in assessing the extent of injury (**Fig. 14.44B**).

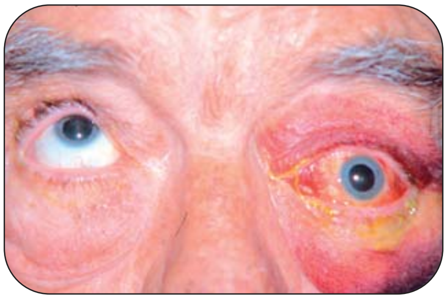

Fig. 14.44A

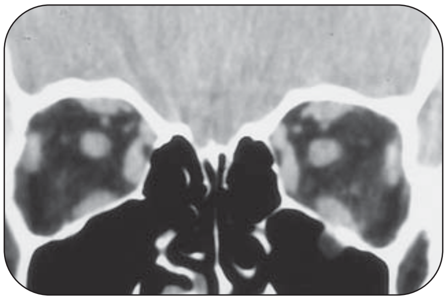

Fig. 14.44B

Look for

- Infraorbital nerve anaesthesia.
- Ocular damage.

Acute myositis

Signs

- Limitation is in the direction of the field of action of the affected muscle.

Look for

- Pain.
- Chemosis.
- Proptosis.

Frozen orbit

Cause

- Severe prolonged idiopathic orbital inflammatory disease (pseudo-tumour).

Signs

- Fibrosis of orbital tissue results in severe ophthalmoplegia (**Fig. 14.45A–D**).

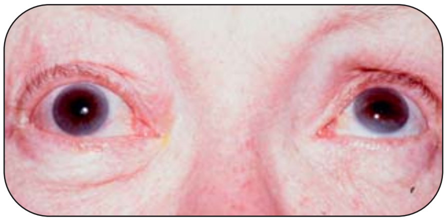

Fig. 14.45A

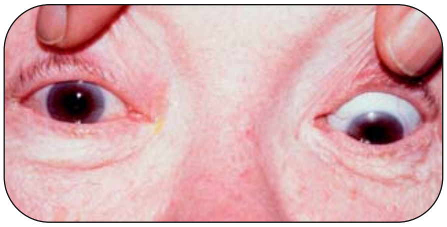

Fig. 14.45B

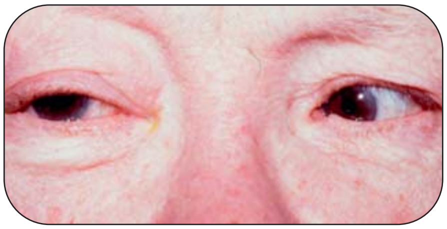

Fig. 14.45C

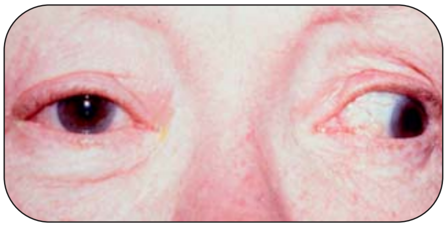

Fig. 14.45D

Chronic progressive external ophthalmoplegia

Signs

- Bilateral ptosis.
- Initially defective upgaze.
- Later generalised symmetric impairment of movement (**Fig. 14.46A** – looking up; **Fig. 14.46B** – looking down; **Fig. 14.46C** – looking right; **Fig. 14.46D** – looking left).

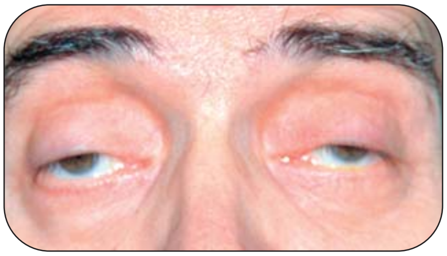

Fig. 14.46A

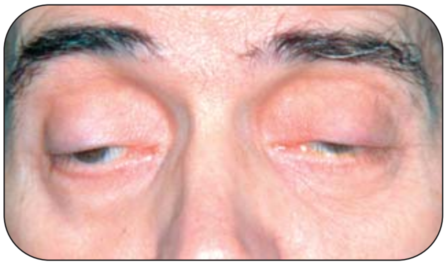

Fig. 14.46B

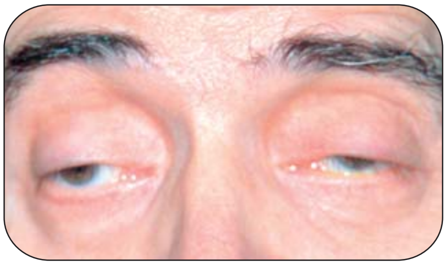

Fig. 14.46C

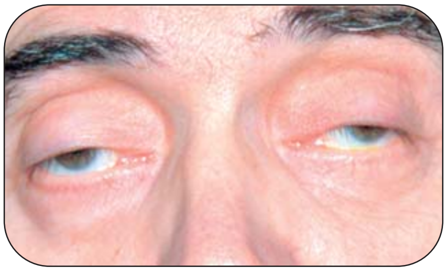

Fig. 14.46D

Look for

a. *Kearns–Sayre syndrome* – pigmentary retinopathy, heart block and ataxia.

b. *Oculopharyngeal dystrophy* – weakness of temporalis and pharyngeal weakness.

Iatrogenic palsies

Causes

- Excessive resection or recession of a vertical or horizontal rectus.
- Slipped muscle during surgery will result in gross underaction.
- Retinal detachment surgery with placement of the explant under the superior or inferior rectus.
- Molteno tube in which the reservoir footplate is placed under the superior rectus.

GAZE PALSIES

Horizontal gaze palsy

Medial longitudinal fasciculus lesion

A medial longitudinal fasciculus (MLF) lesion causes internuclear ophthalmoplegia (INO) which may be unilateral or bilateral.

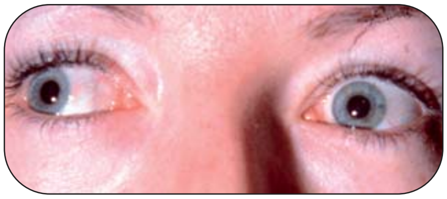

Fig. 14.47A

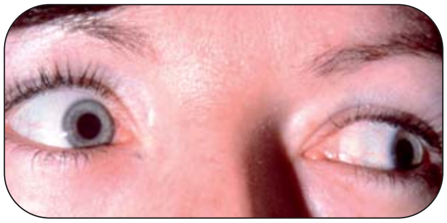

Fig. 14.47B

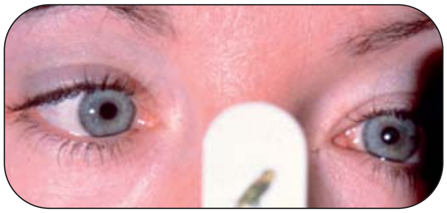

Fig. 14.47C

Signs (left)

- Limitation of left adduction (**Fig. 14.47A**) with ataxic nystagmus of the right eye.
- Normal left abduction (**Fig. 14.47B**).
- Convergence is intact if the lesion is discrete (**Fig. 14.47C**).
- Vertical nystagmus on attempted upgaze.

Causes

- Table 14.3.

Table 14.3 **Causes of internuclear ophthalmoplegia**

- Demyelination (usually bilateral INO)
- Infarction
- Haemorrhage
- Tumours of the brainstem and 4th ventricle
- Infection
- Hydrocephalus
- Progressive supranuclear palsy

Paramedian pontine reticular formation lesion

A paramedian pontine reticular formation (PPRF) lesion causes an ipsilateral gaze palsy with inability to look in the direction of the lesion.

Combined MLF and ipsilateral PPRF (or abducens nucleus) lesion

Signs (left)

- Complete absence of left gaze (**Fig. 14.48A**).
- The only residual movement is abduction of the right eye which exhibits ataxic nystagmus (**Fig. 14.48B**).

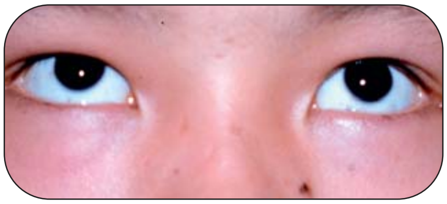

Fig. 14.48A

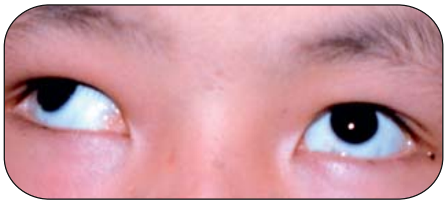

Fig. 14.48B

Bilateral pontine lesions

Bilateral pontine lesions cause total loss of horizontal gaze.

Vertical gaze palsy

Cerebrovascular accidents

Signs

- Midbrain infarction causes complete palsy.
- Multi-infarct syndrome causes incomplete palsy.
- Bilateral lesions of periaqueductal grey cause downgaze palsy with upward tonic deviation of eyes.

Parinaud dorsal midbrain syndrome

Signs

- Restriction of upgaze (**Fig. 14.49A**).
- Convergence paralysis (**Fig. 14.49B**).
- Large pupils with light-near dissociation.
- Lid retraction (Collier sign).
- Convergence-retraction nystagmus.

Causes

- See Table 1.6.

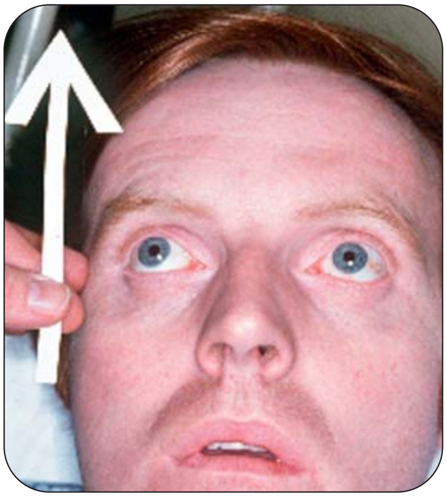

Fig. 14.49A

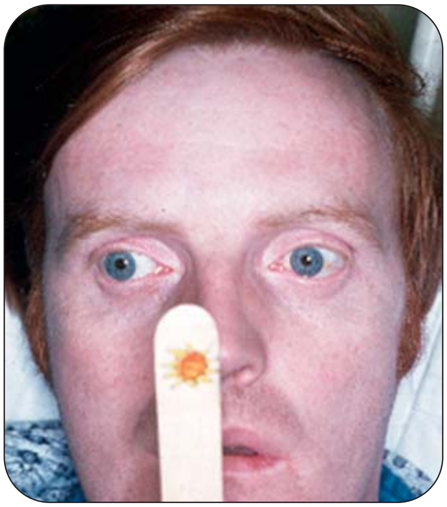

Fig. 14.49B

Progressive supranuclear palsy (Steele–Richardson–Olszewski syndrome)

This is a life-threatening degenerative disorder characterised by severe impairment of voluntary gaze, abnormal posture and tone, pseudo-bulbar palsy and dementia.

NYSTAGMUS

Physiological

Signs (Fig. 14.50)

- Low-amplitude jerk nystagmus in extremes of horizontal gaze with the fast component in the direction of gaze.

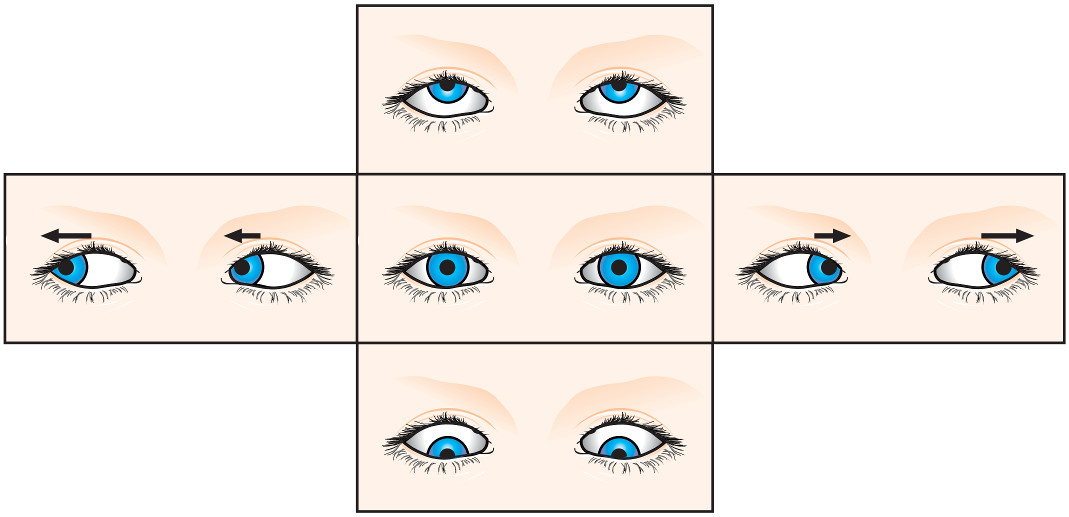

Fig. 14.50

Primary congenital

Presentation

- Within first 6 months of life.

Signs (Fig. 14.51)

- In the primary position there is low-amplitude pendular nystagmus that converts to jerk nystagmus on side gaze.
- In upgaze and downgaze the nystagmus remains in the horizontal plane.

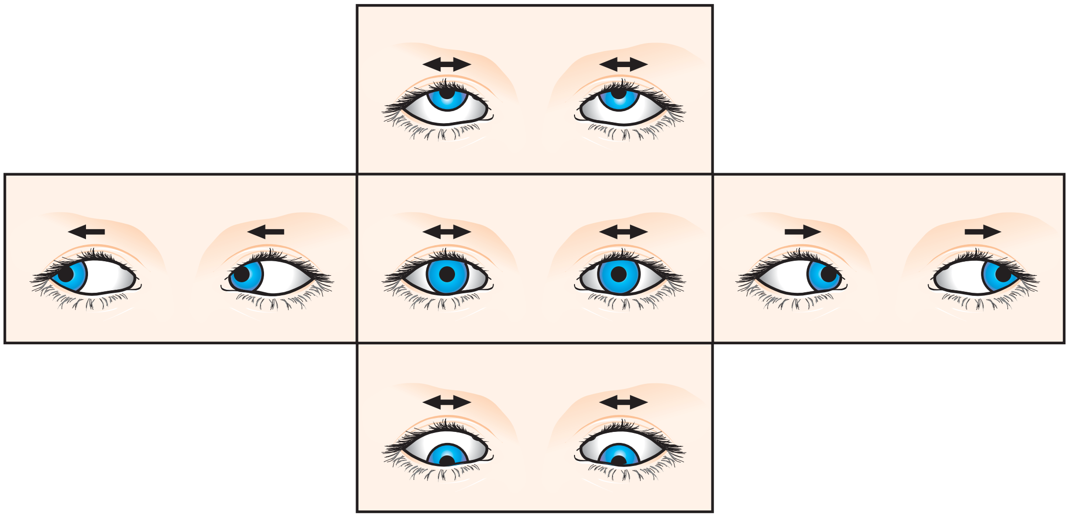

Fig. 14.51

Acquired ocular

Signs

- Pendular nystagmus with an oval trajectory.

Cause

- Severe bilateral impairment of central vision early in life.

Peripheral vestibular

Signs

- Counter-clockwise horizontal rotary nystagmus in all positions of gaze (**Fig. 14.52**).

Causes

- Labrynthitis.
- Ménière disease.
- Middle or inner ear infections.

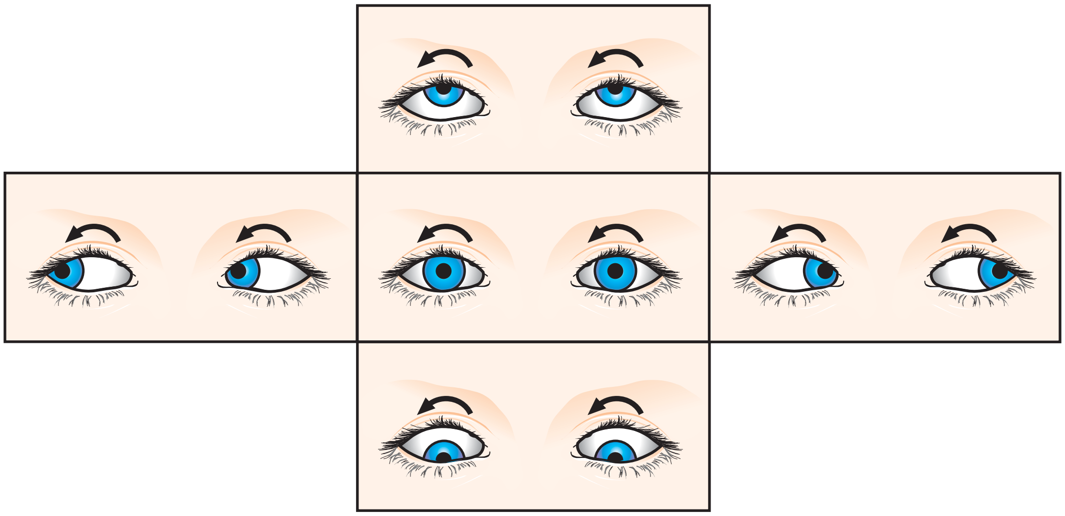

Fig. 14.52

Sidebeat

Signs (Fig. 14.53)

- No nystagmus in the primary position, upgaze and downgaze.
- In right gaze there is right jerk nystagmus.
- In left gaze there is left jerk nystagmus.

Causes

- Neurological disease.

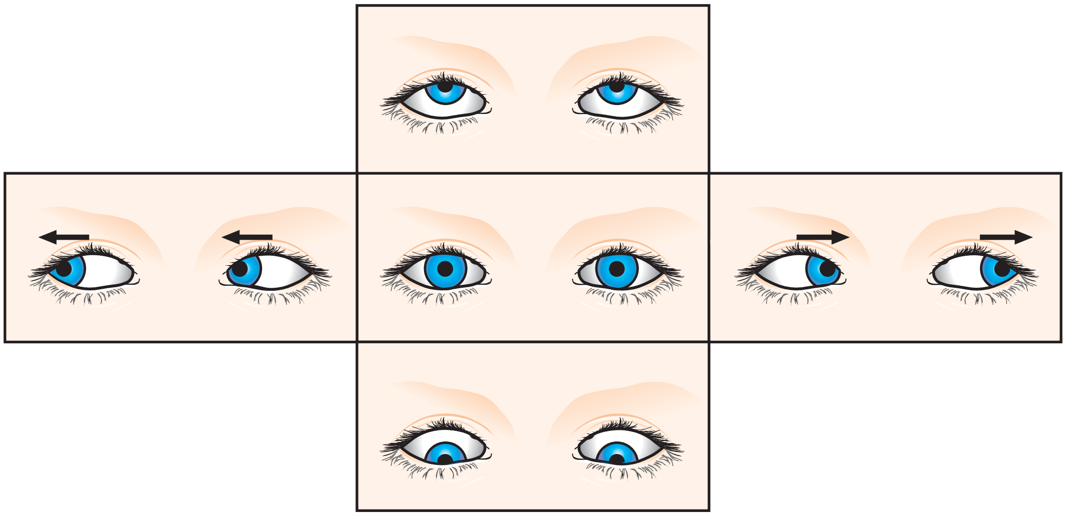

Fig. 14.53

Spasmus nutans

Presentation

- Between 3 and 18 months of age.

Signs

- Low-amplitude high-frequency pendular nystagmus in all positions of gaze (**Fig. 14.54**) often associated with head bobbing.

Causes

- Idiopathic or neurological disease.

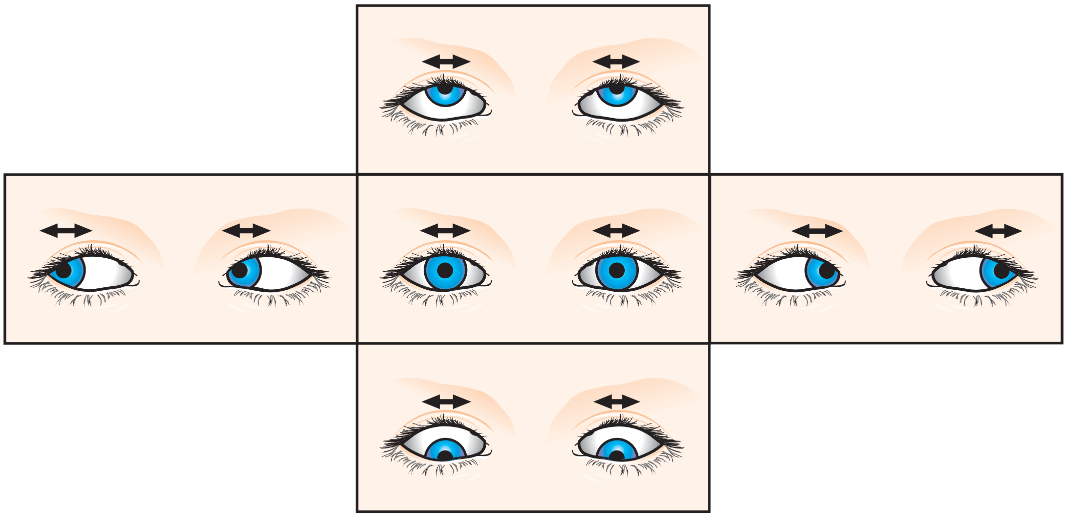

Fig. 14.54

Upbeat

Signs

- Vertical jerk nystagmus with the fast phase beating upwards in all positions of gaze (**Fig. 14.55**).

Causes

- Posterior fossa lesions.
- Drugs.
- Wernicke encephalopathy.

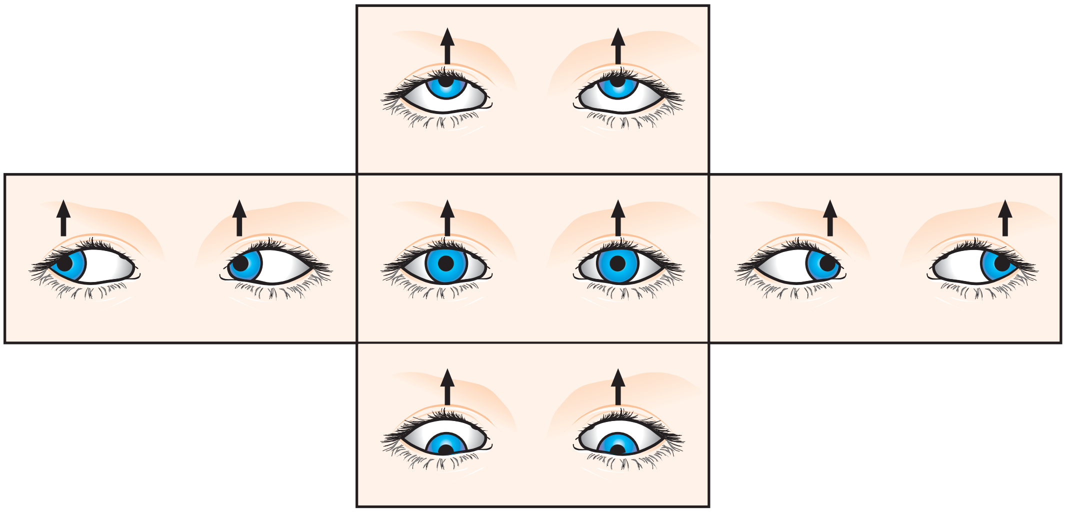

Fig. 14.55

Downbeat

Signs

- Vertical jerk nystagmus with a fast phase beating downwards in all positions of gaze but most pronounced in downgaze (**Fig. 14.56**).

Causes

- Chiari malformation.
- Drugs.
- Acquired neurological disease.

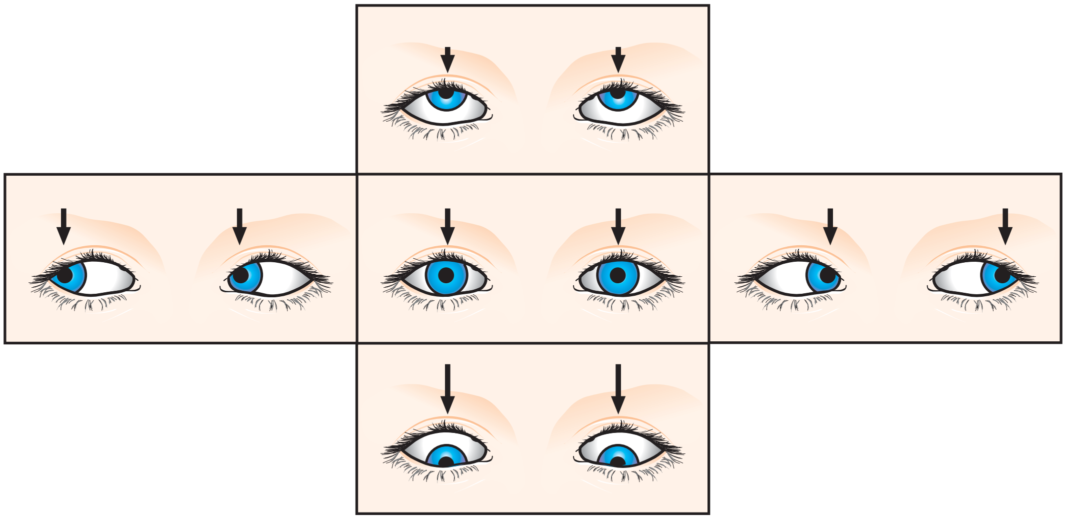

Fig. 14.56

Convergence-retraction

Signs

- On attempted upgaze the eyes converge and retract a few times.

Causes

- See Table 14.3.

Systemic signs

15

FACE **385**

Facial rash **385**
- *Atopic eczema* 385
- *Acne rosacea* 385
- *Sarcoidosis (lupus pernio)* 386
- *Systemic lupus erythematosus* 386
- *Dermatomyositis* 387
- *Tuberous sclerosis (adenoma sebaceum)* 387
- *Xeroderma pigmentosum* 387

Facial pigmentary changes **388**
- *Port wine stain (naevus flammeus)* 388
- *Naevus of Ota (oculodermal melanocytosis)* 388

Lack of facial expression **389**
- *Myotonic dystrophy* 389
- *Myasthenia gravis* 389
- *Systemic sclerosis* 390
- *Parkinsonism* 390
- *Meretoja syndrome* 391
- *Möbius syndrome* 391

Facial spasm **391**
- *Essential blepharospasm* 391
- *Facial hemispasm* 392

Facial hirsutism **392**
- *Cushing syndrome* 392
- *Acromegaly* 393
- *Turner syndrome* 393
- *Porphyria cutanea tarda* 394

Fair complexion and blond hair **394**
- *Oculocutaneous albinism* 394
- *Homocystinuria* 395

Saddle-shaped nose **395**
- *Congenital syphilis* 395
- *Wegener granulomatosis* 395
- *Relapsing polychondritis* 396
- *Leprosy* 396

MOUTH **397**

Ulceration **397**
- *Behçet disease* 397
- *Reiter syndrome* 397
- *Stevens–Johnson syndrome* 397
- *Mucous membrane pemphigoid* 398
- *Pemphigus vulgaris* 398
- *Secondary syphilis* 398

Abnormal dentition **399**
- *Congenital syphilis* 399
- *Osteogenesis imperfecta type 1 and 2* 399
- *Rieger syndrome* 399
- *Dental caries* 399
- *Staining and hypoplasia* 400

High-arched palate **400**
- *Marfan syndrome* 400
- *Pseudoxanthoma elasticum* 400
- *Congenital syphilis* 401
- *Homocystinuria* 401
- *Stickler syndrome* 402

Abnormal tongue **402**
- *Sjögren syndrome* 402
- *Iron deficiency anaemia* 403
- *Acromegaly* 403
- *Multiple endocrine neoplasia type IIb (Sipple syndrome)* 403
- *Rendu–Osler–Weber syndrome* 404

SKULL **404**

Lytic bony lesions **404**
- *Metastatic carcinoma* 404
- *Multiple myeloma* 404
- *Langerhans cell histiocytosis* 405

Large skull **405**
- *Paget disease* 405
- *Acromegaly* 405

HANDS **406**

Long fingers (arachnodactyly) **406**
- *Marfan syndrome* 406
- *Homocystinuria* 406

Short stubby fingers (brachydactyly) **406**
- *Weill–Marchesani syndrome* 406

Other signs **406**
- *Wasting of small muscles* 406
- *Finger clubbing* 407
- *Prayer sign* 407
- *Hyperextensibility of joints* 407
- *Sclerodactyly* 408
- *Raynaud phenomenon* 408
- *Loss of digits* 408
- *Polydactyly* 408
- *Syndactyly* 409

NAILS **409**
- *Pitting* 409
- *Onycholysis* 409
- *Nail dystrophy* 410
- *White nails (leukonychia)* 410
- *Spoon-shaped nails (koilonychia)* 410
- *Subungual splinter haemorrhages* 411
- *Nail-fold infarcts* 411

ARTHRITIS **412**
- *Spinal* 412
- *Peripheral* 412

SKIN **413**
- *Erythema nodosum* 413
- *Vitiligo* 414
- *Localised hypopigmented patches* 414
- *Acanthosis nigricans* 414
- *Generalised hyperpigmentation* 415
- *Vesiculobullous dermatoses* 415
- *Pyoderma gangrenosum* 415
- *Cutaneous bleeding* 416
- *Thin hyperelastic skin* 416
- *Vasculitis* 416
- *Telangiectasia* 417
- *Livedo reticularis* 417
- *Necrobiosis lipoidica* 417
- *Acquired ichthyosis* 418
- *Multiple tumours* 418

GENITALIA **418**
- *Urethritis* 418
- *Ulceration* 419
- *Balanitis and phimosis* 420

FACE

Facial rash

Atopic eczema

Signs

- Dry, itchy, erythematous thickening of facial skin (**Fig. 15.1**).

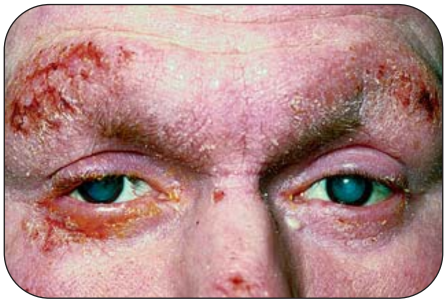

Fig. 15.1

Look for

- Staphylococcal blepharitis.
- Vernal and atopic keratoconjunctivitis.
- Early-onset shield-like cataract.
- Keratoconus.
- Retinal detachment.

Acne rosacea

Signs

- Erythema and papules on the glabella, cheeks, nose and chin (**Fig. 15.2**).

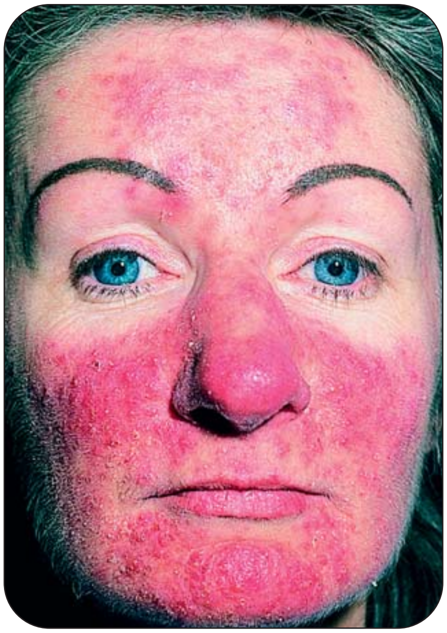

Fig. 15.2

Look for

- Chronic posterior blepharitis.
- Recurrent chalazion.
- Peripheral keratitis.

Sarcoidosis (lupus pernio)

Signs

- Violaceous lesions with a predilection for the nose (**Fig. 15.3**).

Look for

- Uveitis.
- Retinal periphlebitis.
- Fundus granulomas.
- Dry eye.
- Conjunctival granulomas.

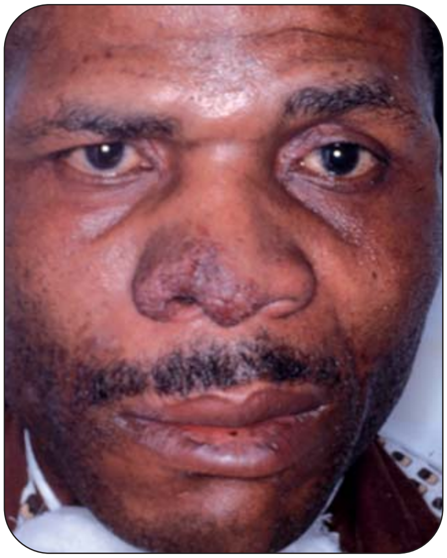

Fig. 15.3

Systemic lupus erythematosus

Signs

- Erythematous rash involving the cheeks and bridge of the nose with a ‘butterfly’ distribution (**Fig. 15.4**).

Look for

- Dry eye.
- Peripheral ulcerative keratitis.
- Scleritis.
- Retinopathy.

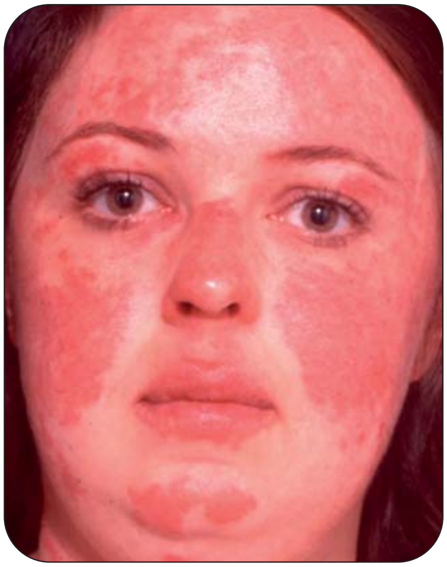

Fig. 15.4

Dermatomyositis

Signs

- Extensive erythematous rash (**Fig. 15.5**).

Look for

- Violaceous (heliotrope) eyelid rash and periorbital oedema.
- Dry eye.
- Scleritis.
- Retinopathy.

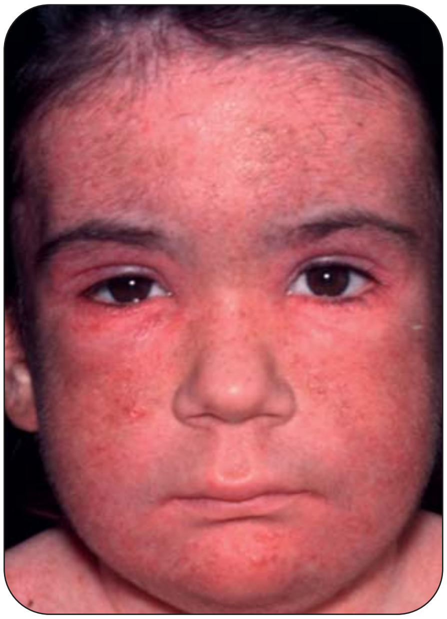

Fig. 15.5

Tuberous sclerosis (adenoma sebaceum)

Signs

- Red papules on the cheeks and nose (**Fig. 15.6**).

Look for

- Fundus astrocytomas.

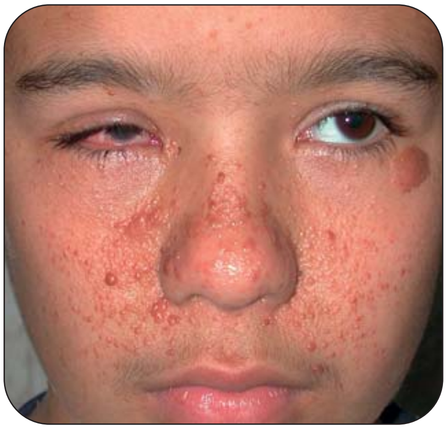

Fig. 15.6

Xeroderma pigmentosum

Signs

- Bird-like facies, and cutaneous atrophy, scaling and pigmentation with a propensity to malignant change (**Fig. 15.7**).

Look for

- Eyelid and conjunctival malignancies.
- Dry eye.

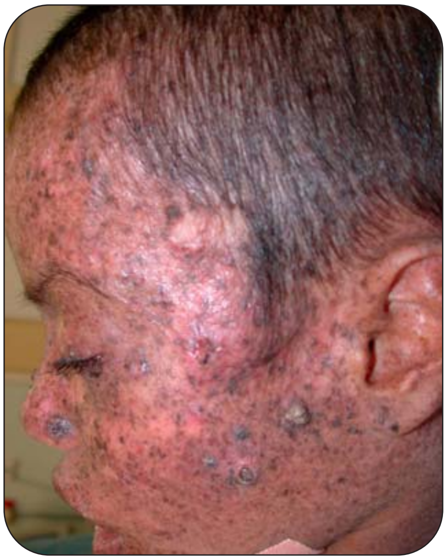

Fig. 15.7

Facial pigmentary changes

Port wine stain (naevus flammeus)

Signs

- Sharply demarcated, usually unilateral, soft, purple patch which occurs in patients with Sturge–Weber syndrome (**Fig. 15.8**).

Look for

- Episcleral haemangioma.
- Diffuse choroidal haemangioma.
- Glaucoma.
- Heterochromia iridis.

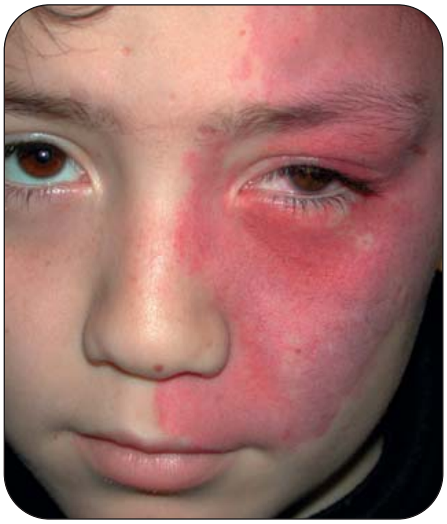

Fig. 15.8

Naevus of Ota (oculodermal melanocytosis)

Signs

- Unilateral hyperpigmentation of deep facial skin, most frequently in the distribution of the 1st and 2nd divisions of the trigeminal nerve (**Fig. 15.9**).

Look for

- Episcleral hyperpigmentation.
- Heterochromia iridis.
- Iris mammillations.
- Trabecular hyperpigmentation and glaucoma.
- Fundus hyperpigmentation.
- Uveal melanoma.

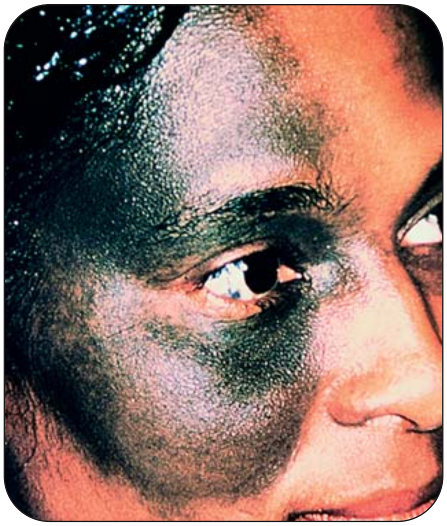

Fig. 15.9

Lack of facial expression

Myotonic dystrophy

Signs

- Bilateral facial wasting with hollow cheeks, sagging jaw and ptosis (**Fig. 15.10**).

Look for

- Early-onset shield-like cataract.
- Light-near dissociation of pupils.
- Mild pigmentary retinopathy.

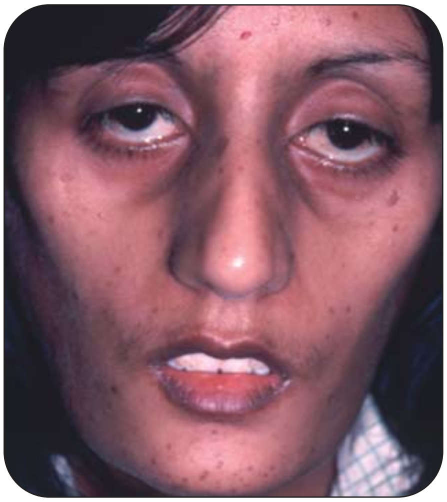

Fig. 15.10

Myasthenia gravis

Signs

- Myasthenic involvement of muscles of facial expression and ptosis (**Fig. 15.11**).

Look for

- Ophthalmoplegia, particularly involving elevation.

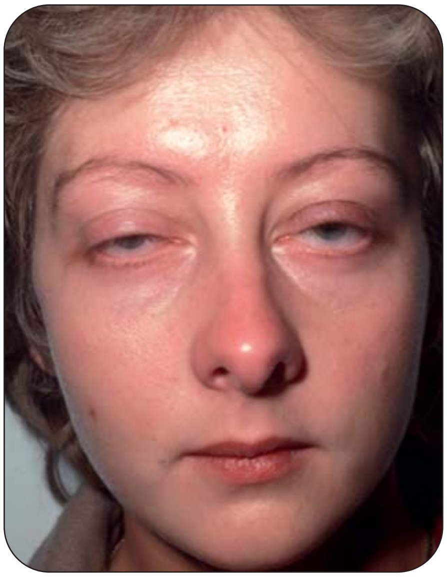

Fig. 15.11

Systemic sclerosis

Signs

- Shiny tight facial skin due to subcutaneous fibrotic changes, nasal deformity and vertical furrowing of perioral skin (**Fig. 15.12**).

Look for

- Eyelid scarring.
- Dry eye.
- Retinopathy.

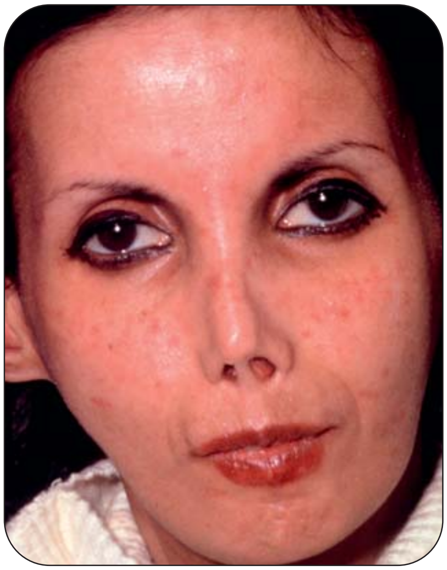

Fig. 15.12

Parkinsonism

Signs

- Sweaty and greasy face with mild lid retraction (**Fig. 15.13**).

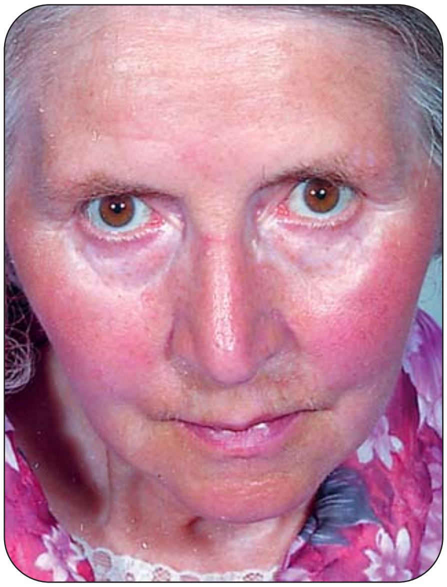

Fig. 15.13

Meretoja syndrome

Signs

- Bilateral facial palsy (**Fig. 15.14**).

Look for

- Corneal lattice dystrophy type 2.

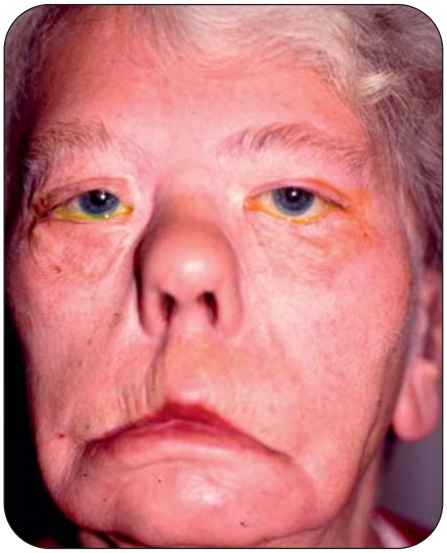

Fig. 15.14

Möbius syndrome

Signs

- Bilateral facial paresis with the mouth constantly held open (**Fig. 15.15**).

Look for

- Esotropia.
- Limitation of abduction.
- Horizontal gaze palsy.

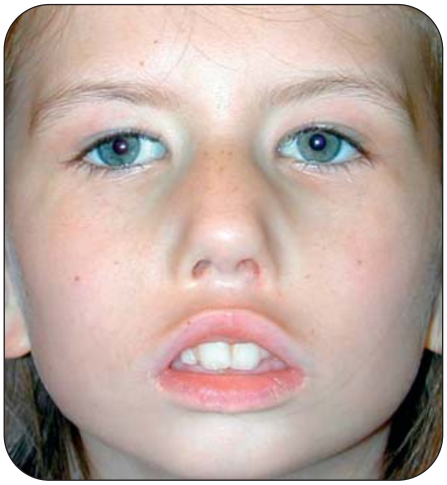

Fig. 15.15

Facial spasm

Essential blepharospasm

Signs

- Periodic bilateral involuntary spasm of the orbicularis oculi and upper facial muscles temporally rendering the patients functionally blind (**Fig. 15.16**).

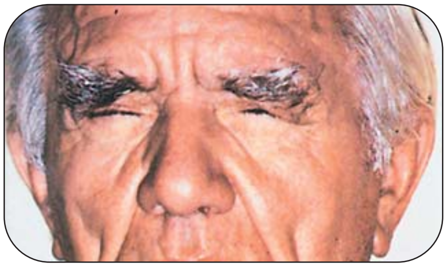

Fig. 15.16

Facial hemispasm

Signs

- Brief unilateral spasm of the orbicularis oculi and facial muscles (**Fig. 15.17**).

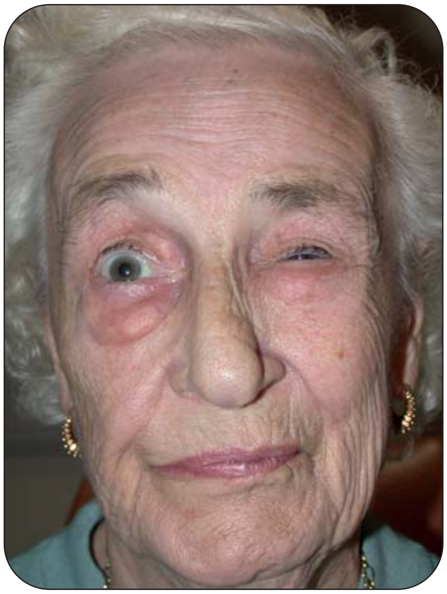

Fig. 15.17

Facial hirsutism

Cushing syndrome

Signs

- Also moon face and hyperpigmentation (**Fig. 15.18**)

Look for

- Bitemporal hemianopia due to adenoma (Cushing disease).
- Cataract.

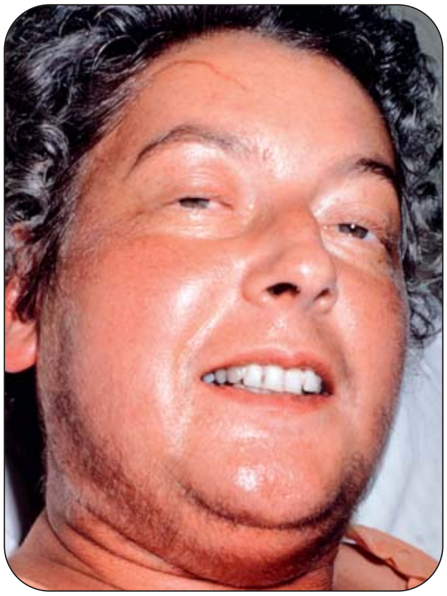

Fig. 15.18

Acromegaly

Signs

- Coarse facies (**Fig. 15.19**).

Look for

- Bitemporal hemianopia due to a pituitary adenoma.
- Optic atrophy.
- Angioid streaks.

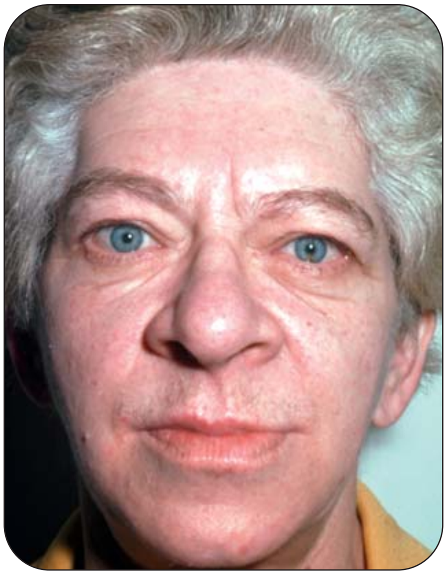

Fig. 15.19

Turner syndrome

Signs

- Webbed neck (**Fig. 15.20**).

Look for

- Keratoconus.
- Blue sclera.
- Cataract.

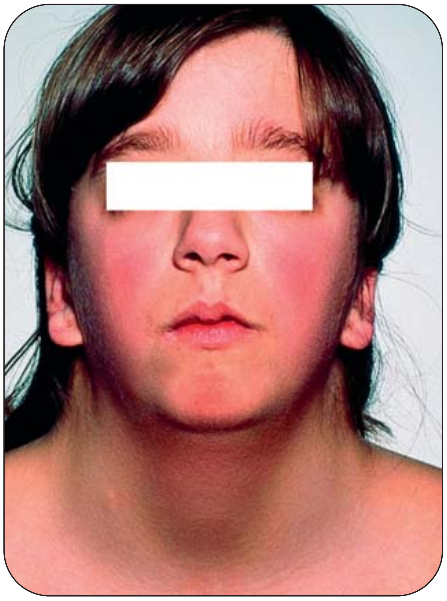

Fig. 15.20

Porphyria cutanea tarda

Signs

- Bullous skin eruptions and scarring (**Fig. 15.21**).

Look for

- Cicatrising conjunctivitis.
- Scleritis.

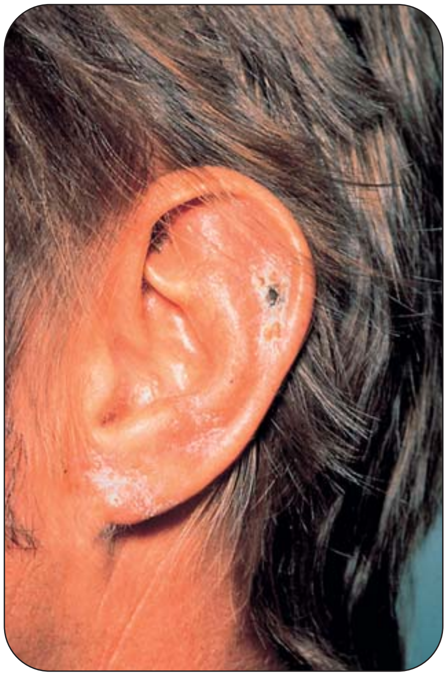

Fig. 15.21

Fair complexion and blond hair

Oculocutaneous albinism

Signs

- Pale skin and blond hair (**Fig. 15.22**).

Look for

- Nystagmus.
- Iris transillumination.
- Pale fundus and foveal hypoplasia.
- Anomalous chiasmal nerve fibre pathway.

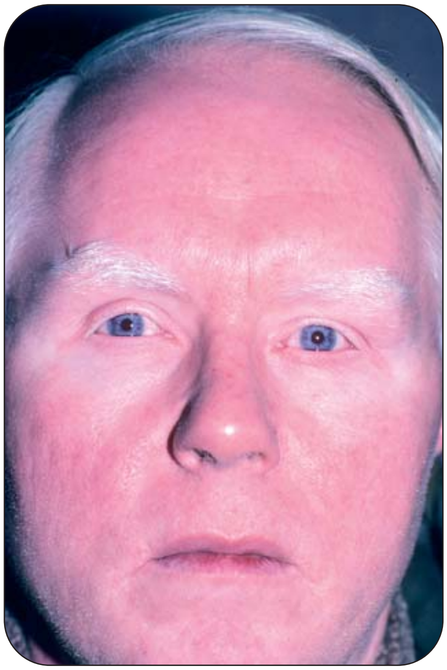

Fig. 15.22

Homocystinuria

Signs

- Coarse fair hair and malar flush (see Fig. 10.16).

Look for

- Ectopia lentis.
- Myopia.
- Retinal detachment.

Saddle-shaped nose

Congenital syphilis

Pathogenesis

- Osteitis of the nasal bone (**Fig. 15.23**).

Look for

- Interstitial keratitis.
- Pigmentary retinopathy.

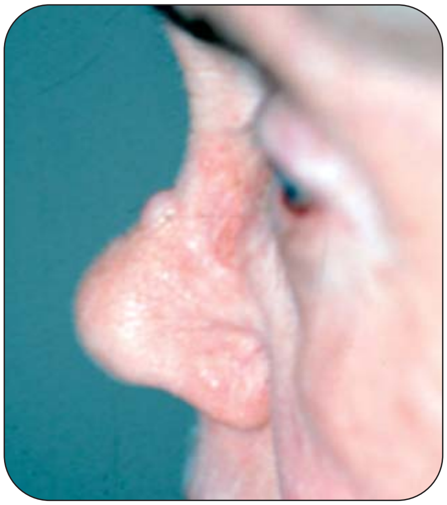

Fig. 15.23

Wegener granulomatosis

Pathogenesis

- Necrotising granulomatous vasculitis involving the nose (**Fig. 15.24**).

Look for

- Scleritis.
- Peripheral ulcerative keratitis.
- Orbital involvement.
- Nasolacrimal duct obstruction.
- Retinopathy.

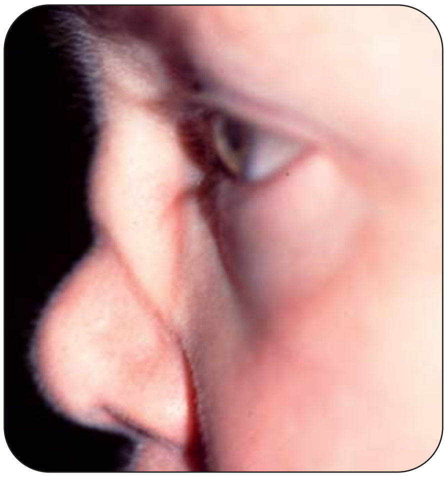

Fig. 15.24

Relapsing polychondritis

Pathogenesis

- Recurrent inflammatory swelling, destruction and collapse of nasal cartilage (**Fig. 15.25**).

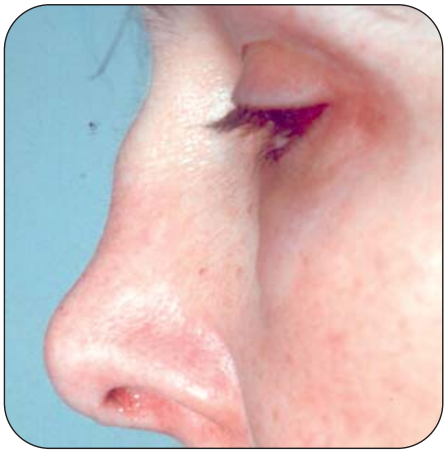

Fig. 15.25

Look for

- Scleritis.
- Anterior uveitis.

Leprosy

Pathogenesis

- Destruction of the nasal bone and cartilage (**Fig. 15.26**).

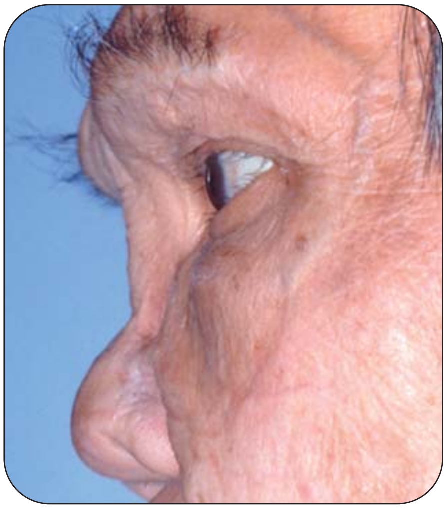

Fig. 15.26

Look for

- Madarosis.
- Anterior uveitis.
- Miosis.
- Iris atrophy.
- Keratitis.
- Scleritis.

MOUTH

Ulceration

Behçet disease

Signs

- Recurrent, painful aphthous stomatitis which may involve the buccal mucosa and the tongue (**Fig. 15.27**).

Look for

- Anterior uveitis with hypopyon.
- Panuveitis.
- Retinal vasculitis.

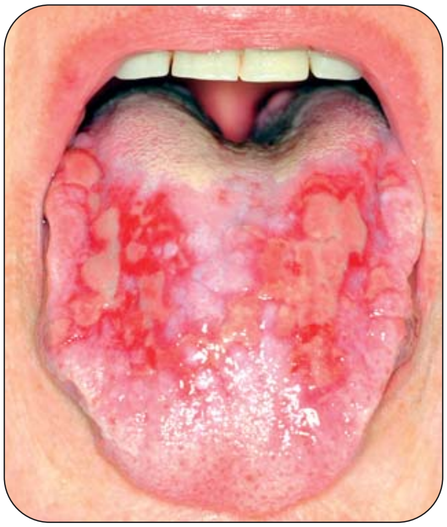

Fig. 15.27

Reiter syndrome

Signs

- Transient painless ulceration (**Fig. 15.28**).

Look for

- Conjunctivitis.
- Acute anterior uveitis.
- Keratitis.

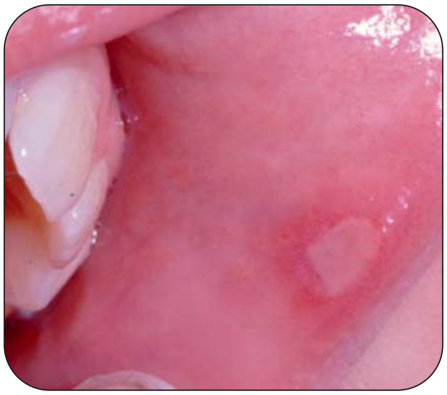

Fig. 15.28

Stevens–Johnson syndrome

Signs

- Mucosal bullae and erosions associated with haemorrhagic crusting of the lips (**Fig. 15.29**).

Look for

- Cicatrising conjunctivitis.

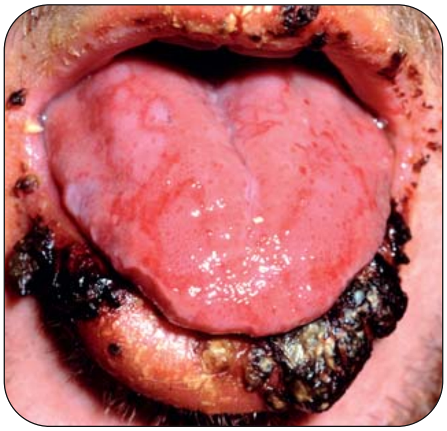

Fig. 15.29

Mucous membrane pemphigoid

Signs

- Mucosal blisters and superficial erosions (**Fig. 15.30**).

Look for

- Cicatrising conjunctivitis.

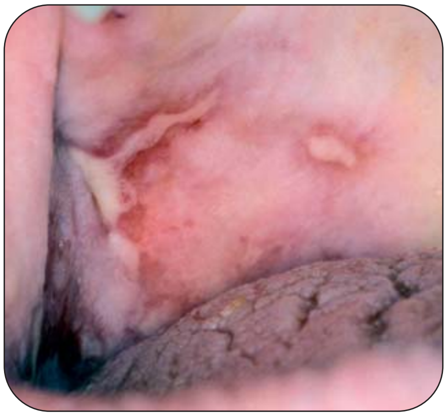

Fig. 15.30

Pemphigus vulgaris

Signs

- Mucosal blisters and erosions (**Fig. 15.31**).

Look for

- Cicatrising conjunctivitis.

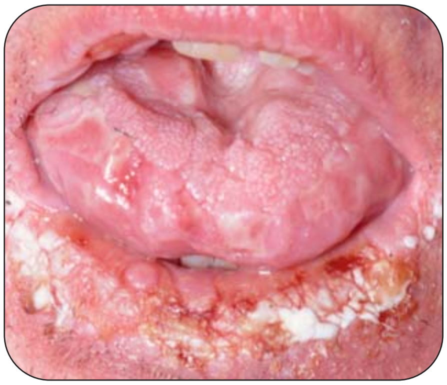

Fig. 15.31

Secondary syphilis

Signs

- 'Snail-track' ulcers (**Fig. 15.32**).

Look for

- Madarosis.
- Anterior uveitis.
- Interstitial keratitis.
- Acute posterior placoid chorioretinitis.
- Neuroretinitis.
- Optic neuritis.

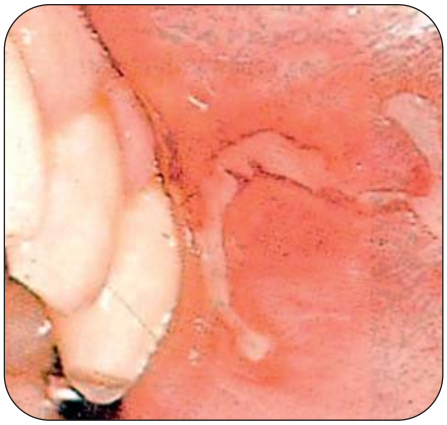

Fig. 15.32

Abnormal dentition

Congenital syphilis

Signs

- Mulberry molars (imperfectly formed cusps) and malformed incisors (Hutchinson teeth – **Fig. 15.33**).

Look for

- Interstitial keratitis.
- Pigmentary retinopathy.

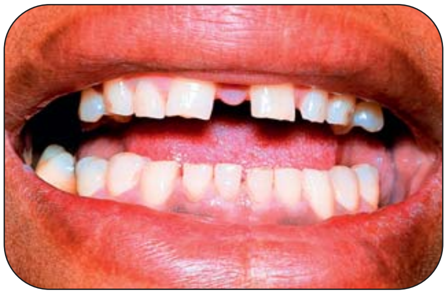

Fig. 15.33

Osteogenesis imperfecta type 1 and 2

Signs

- Small teeth (dentiogenesis imperfecta – **Fig. 15.34**).

Look for

- Blue sclera.

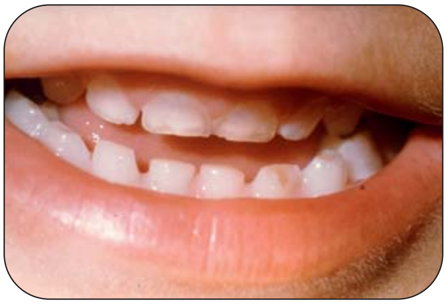

Fig. 15.34

Rieger syndrome

Signs

- Small teeth (microdontia) which are also fewer in number (hypodontia – **Fig. 15.35**).

Look for

- Axenfeld–Rieger syndrome (see Figs 7.22, 7.23, 8.71).

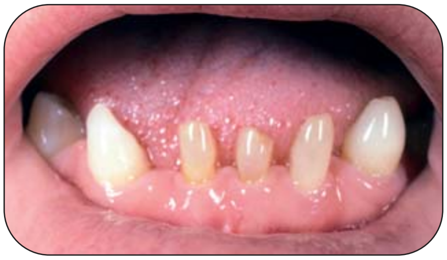

Fig. 15.35

Dental caries

Signs

- Severe caries (**Fig. 15.36**).

Look for

- Sjögren syndrome.

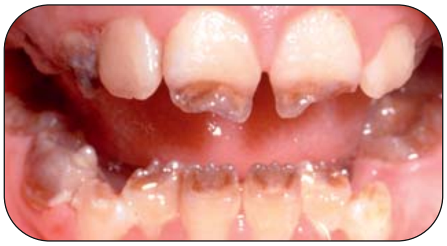

Fig. 15.36

Staining and hypoplasia

Signs

- Yellow discoloration of abnormally small teeth (**Fig. 15.37**) resulting from systemic tetracycline administered to children under the age of 12 years, or to pregnant or breast-feeding women.

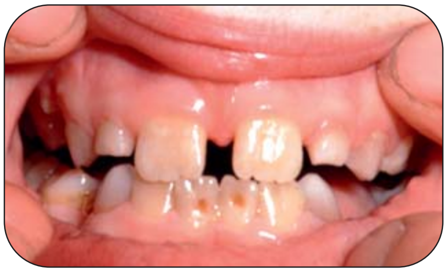

Fig. 15.37

High-arched palate

Marfan syndrome

Signs

- Gothic (cathedral roof) palate (**Fig. 15.38**).

Look for

- Ectopia lentis.
- Myopia.
- Retinal detachment.
- Keratoconus.
- Angle anomaly and glaucoma.

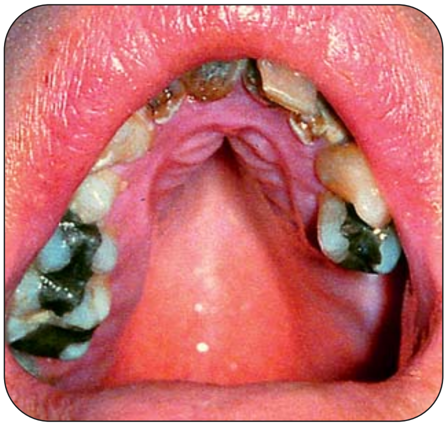

Fig. 15.38

Pseudoxanthoma elasticum

Signs

- Also characteristic skin changes (**Fig. 15.39**).

Look for

- Angioid streaks.
- Blue sclera.

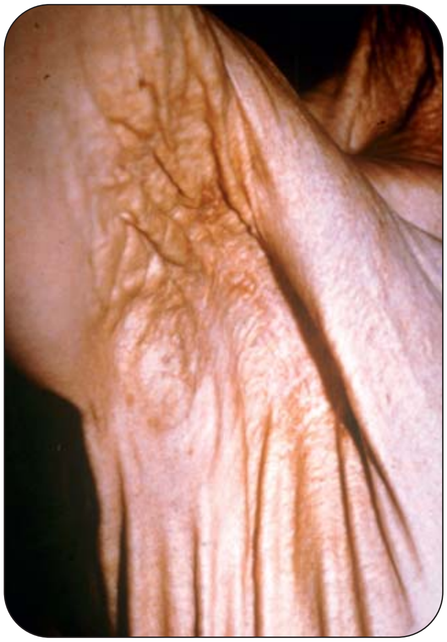

Fig. 15.39

Congenital syphilis

Signs

- Also sabre tibiae from periostitis (**Fig. 15.40**).

Look for

- Interstitial keratitis.
- Pigmentary retinopathy.

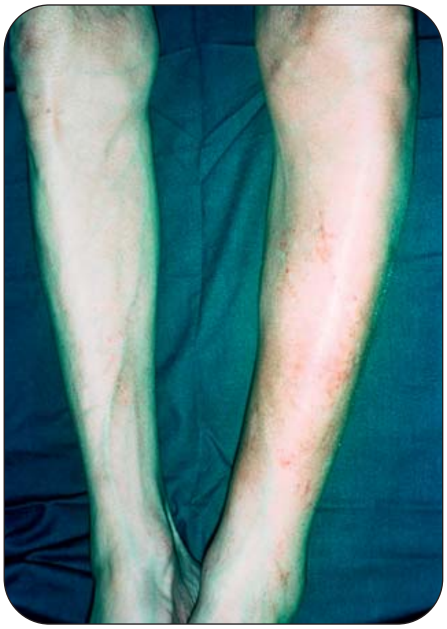

Fig. 15.40

Homocystinuria

Signs

- Also tendency to thrombosis of veins and arteries (**Fig. 15.41**).

Look for

- Ectopia lentis.
- Myopia.
- Retinal detachment.

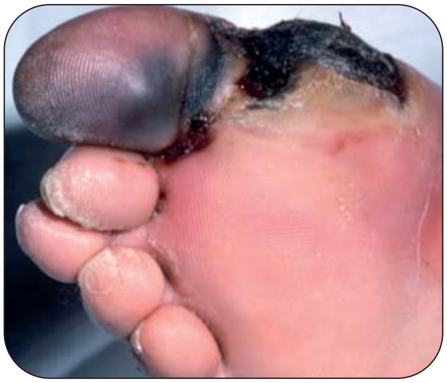

Fig. 15.41

Stickler syndrome

Signs

- Mid-facial flattening with depressed nasal bridge and short nose (**Fig. 15.42**).

Look for

- Congenital non-progressive high myopia.
- Vitreoretinopathy and retinal detachment.
- Ectopia lentis.
- Cataract.
- Glaucoma.

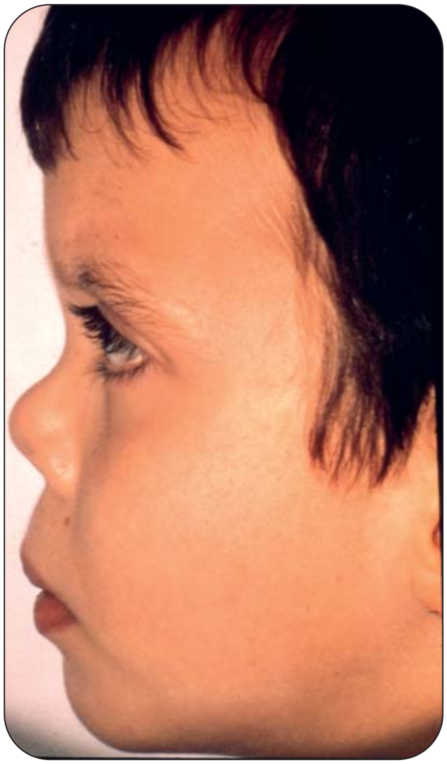

Fig. 15.42

Abnormal tongue

Sjögren syndrome

Signs

- Dry tongue (**Fig. 15.43**).

Look for

- Dry eyes.
- Adie pupil (rare).

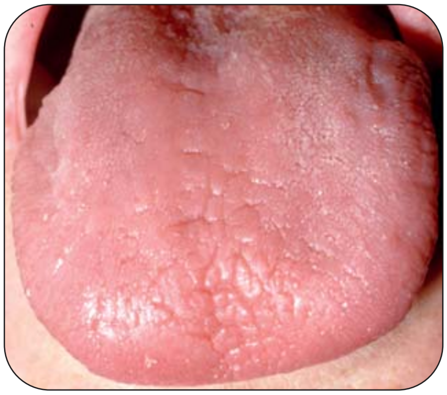

Fig. 15.43

Iron deficiency anaemia

Signs

- Pallor with loss of papillae (**Fig. 15.44**).

Look for

- Retinopathy.

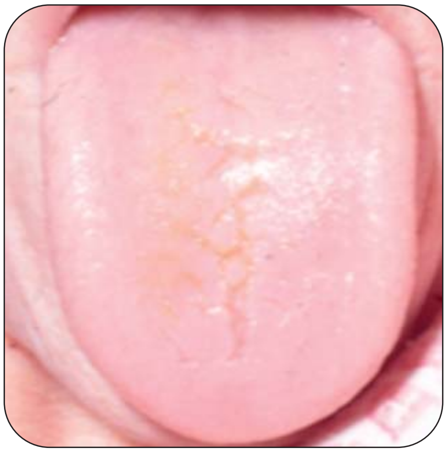

Fig. 15.44

Acromegaly

Signs

- Large tongue (macroglossia – **Fig. 15.45**).

Look for

- Bitemporal hemianopia due to a pituitary adenoma.
- Optic atrophy.
- Angioid streaks.

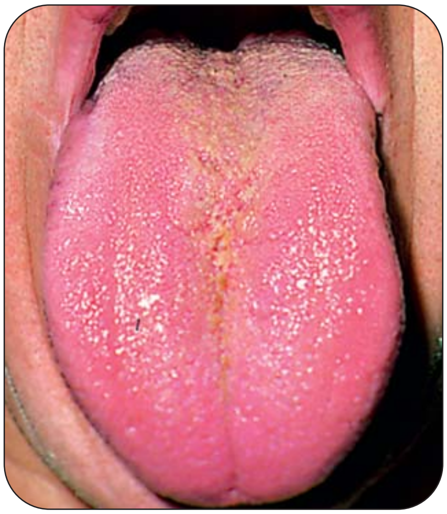

Fig. 15.45

Multiple endocrine neoplasia type IIb (Sipple syndrome)

Signs

- Thickening due to multiple neuromas (**Fig. 15.46**).

Look for

- Eyelid neurofibroma.
- Prominent corneal nerves (see Figs 5.28 and 5.29).
- Conjunctival neuromas.
- Poor pupillary dilatation.

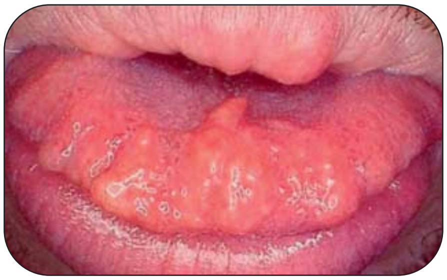

Fig. 15.46

Rendu–Osler–Weber syndrome

Signs

- Telangiectasia (**Fig. 15.47**).

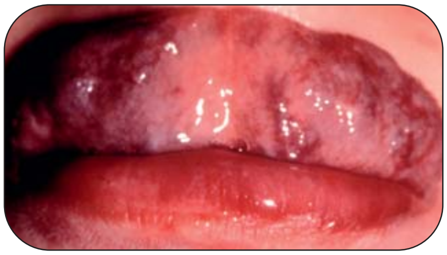

Fig. 15.47

Look for

- Conjunctival telangiectasia (see Fig. 3.61).
- Retinal haemorrhages.

SKULL

Lytic bony lesions

Metastatic carcinoma

Signs

- Multiple lytic lesions (**Fig. 15.48**) most frequently from breast, bronchus and prostate.

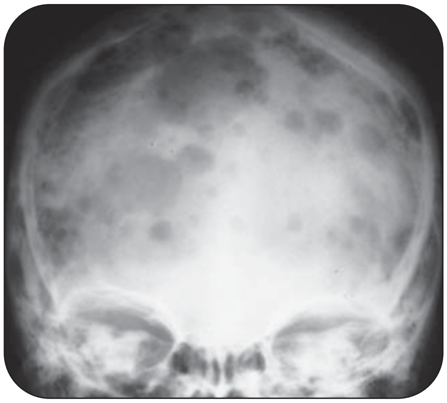

Fig. 15.48

Look for

- Uveal metastases but not from prostate.

Multiple myeloma

Signs

- Multiple lytic lesions which are larger than in metastatic carcinoma (**Fig. 15.49**).

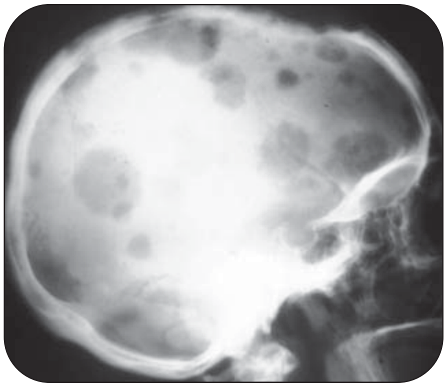

Fig. 15.49

Look for

- Retinopathy.

Langerhans cell histiocytosis

Signs

- Large solitary lytic lesion (**Fig. 15.50**).

Look for

- Unilateral or bilateral proptosis.

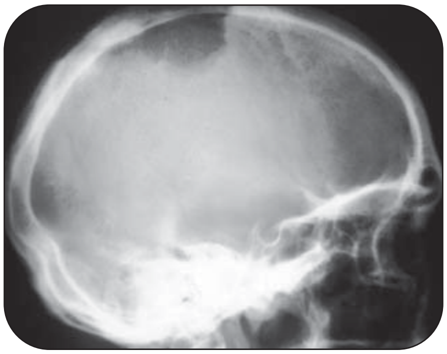

Fig. 15.50

Large skull

Paget disease

Signs

- Skull enlargement associated with osteosclerotic expansion (**Fig. 15.51**).

Look for

- Optic atrophy.
- Angioid streaks.
- Ocular motor nerve palsies.
- Proptosis.

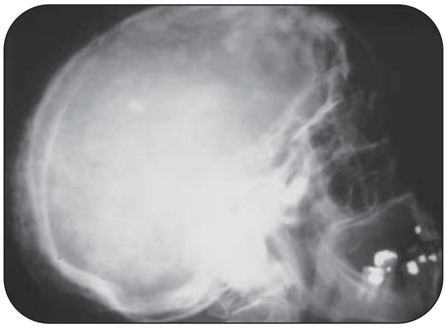

Fig. 15.51

Acromegaly

Signs

- Enlargement of the skull and mandible (**Fig. 15.52**).

Look for

- Bitemporal hemianopia due to a pituitary adenoma.
- Optic atrophy.
- Angioid streaks.

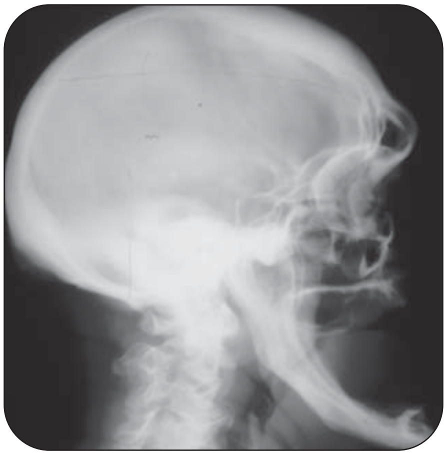

Fig. 15.52

HANDS

Long fingers (arachnodactyly)

Marfan syndrome

Signs

- Long spider-like fingers (**Fig. 15.53**) and toes.

Look for

- Bilateral superior ectopia lentis (see Fig. 10.10).
- Retinal detachment.
- Angle anomaly and glaucoma.

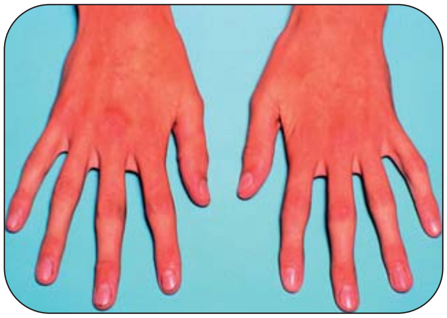

Fig. 15.53

Homocystinuria

Look for

- Ectopia lentis.
- Myopia.
- Retinal detachment.

Short stubby fingers (brachydactyly)

Weill–Marchesani syndrome

Signs

- Spade-like hands with short stubby fingers (**Fig. 15.54**).

Look for

- Bilateral inferior ectopia lentis.
- Pupil block glaucoma.

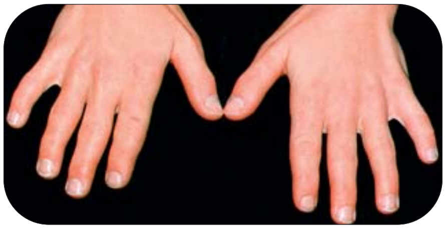

Fig. 15.54

Other signs

Wasting of small muscles (Fig. 15.55)

Look for

- Pancoast tumour of the bronchus.
- Syringomyelia.
- Charcot–Marie–Tooth disease.

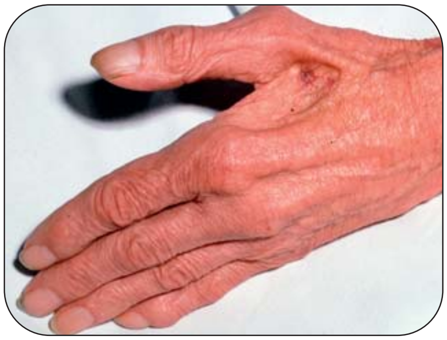

Fig. 15.55

Finger clubbing (Fig. 15.56)

Look for

- Bronchial carcinoma.
- Thyroid disease (acropachy).
- Bacterial endocarditis.
- Crohn disease.
- Ulcerative colitis.
- Whipple disease.

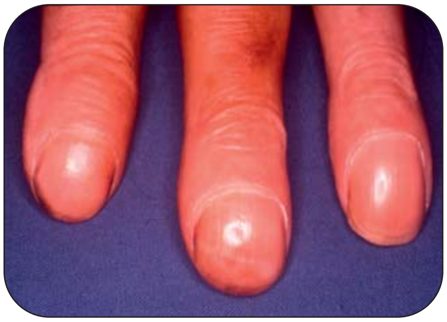

Fig. 15.56

Prayer sign

Signs

- Inability to press the palms of the hands together (**Fig. 15.57**)

Look for

- Stenosing tendonitis in type 1 diabetes.

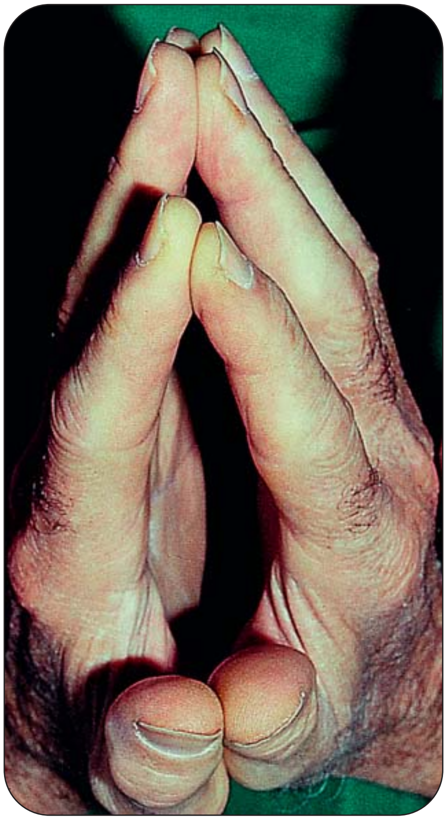

Fig. 15.57

Hyperextensibility of joints (Fig. 15.58)

Look for

- Ehlers–Danlos syndrome.

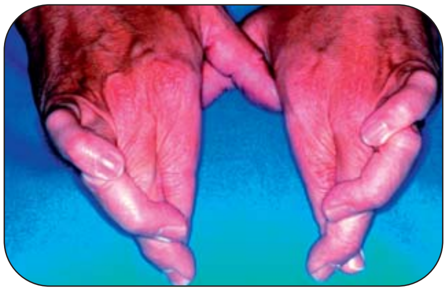

Fig. 15.58

Sclerodactyly

Signs

- Shiny, creaseless, flexed tapered fingers with loss of finger pulps (**Fig. 15.59**).

Look for

- Scleroderma.
- Dermatomyositis.

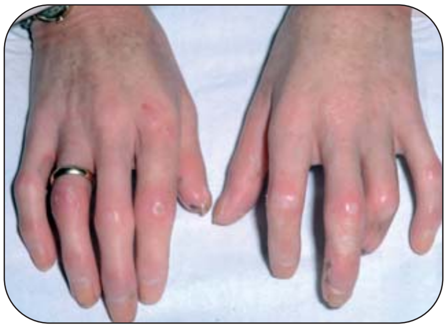

Fig. 15.59

Raynaud phenomenon

Signs

- Intermittent, bilateral vasospasm following exposure to cold that may progress to gangrene in severe cases (**Fig. 15.60**).

Look for

- Scleroderma.
- Rheumatoid arthritis.
- Systemic lupus erythematosus.
- Dermatomyositis.
- Sjögren syndrome.
- Waldenström macroglobulinaemia.

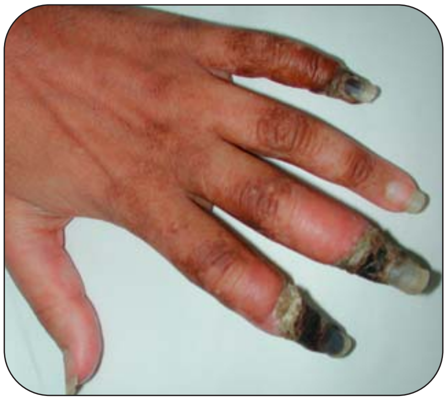

Fig. 15.60

Loss of digits (Fig. 15.61)

Look for

- Leprosy.

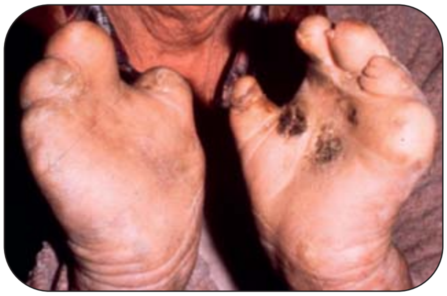

Fig. 15.61

Polydactyly (Fig. 15.62)

Look for

- Bardet–Biedl syndrome.

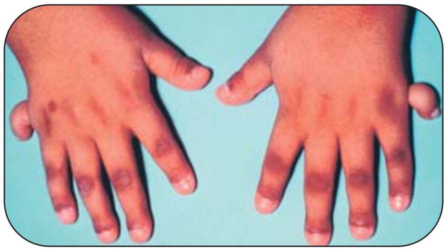

Fig. 15.62

Syndactyly (Fig. 15.63)

Look for

- Apert syndrome.

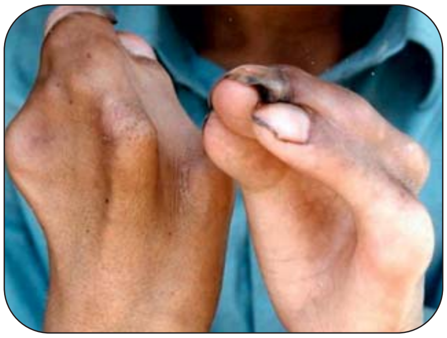

Fig. 15.63

NAILS

Pitting (Fig. 15.64)

Look for

- Psoriasis and psoriatic arthritis.

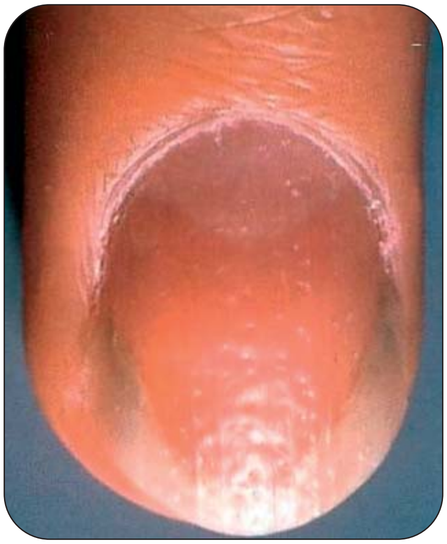

Fig. 15.64

Onycholysis (Fig. 15.65)

Look for

- Psoriasis and psoriatic arthritis.

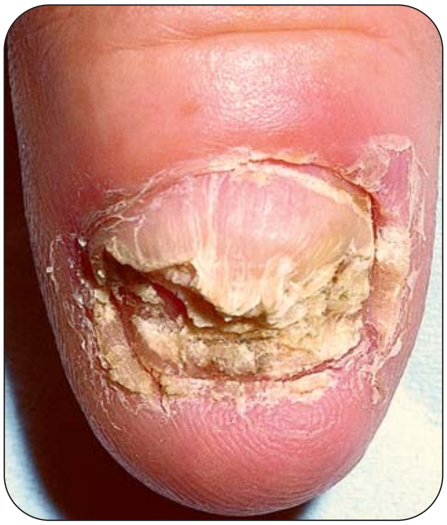

Fig. 15.65

Nail dystrophy (Fig. 15.66)

Look for

- Epidermolysis bullosa.

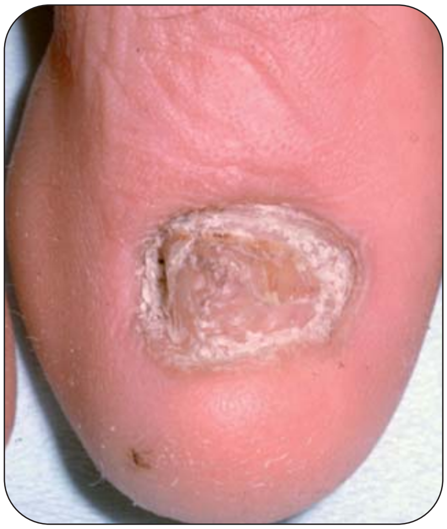

Fig. 15.66

White nails (leukonychia) (Fig. 15.67)

Look for

- Liver disease.

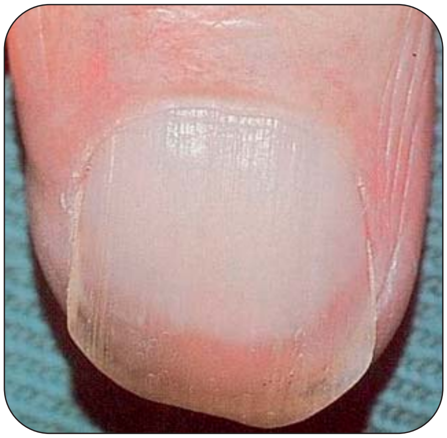

Fig. 15.67

Spoon-shaped nails (koilonychia) (Fig. 15.68)

Look for

- Iron deficiency anaemia.

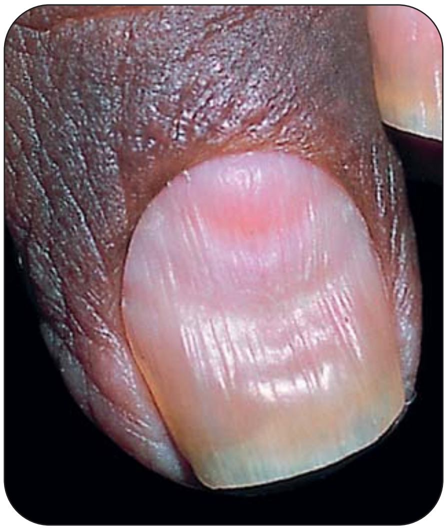

Fig. 15.68

Subungual splinter haemorrhages (Fig. 15.69)

Look for

- Bacterial endocarditis.

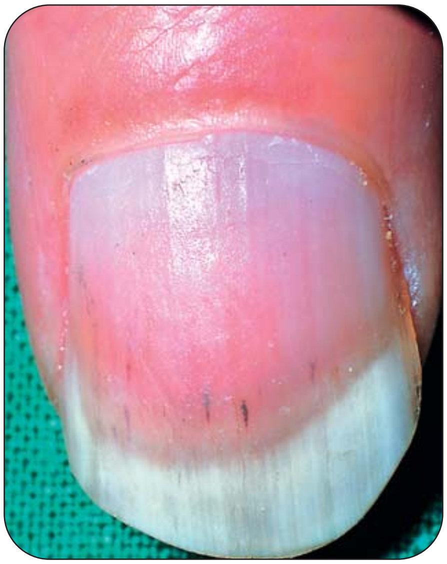

Fig. 15.69

Nail-fold infarcts (Fig. 15.70)

Look for

- Rheumatoid arthritis.
- Systemic lupus erythematosus.
- Dermatomyositis.

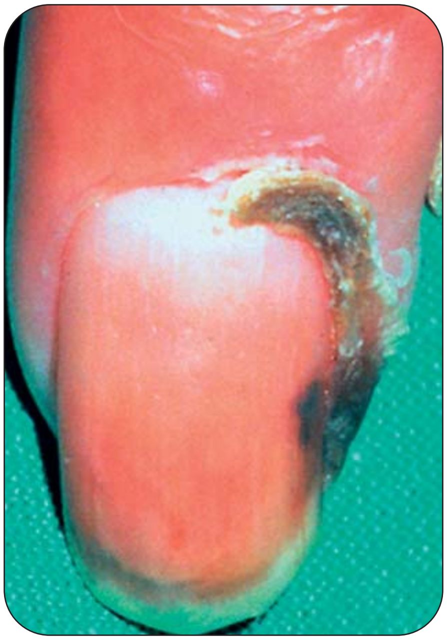

Fig. 15.70

ARTHRITIS

Spinal

Look for

a. *Ankylosing spondylitis* – rigid neck is typical (**Fig. 15.71**).

b. *Reiter syndrome* (common).

c. *Psoriatic arthritis* (uncommon).

d. *Ulcerative colitis* (common).

e. *Crohn disease* (uncommon).

f. *Whipple disease* (uncommon).

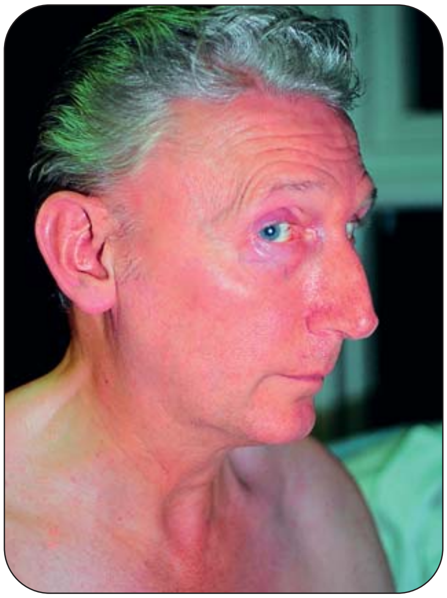

Fig. 15.71

Peripheral

Look for

a. *Rheumatoid arthritis* – symmetrical arthritis with ulnar deviation (**Fig. 15.72**).

b. *Systemic lupus erythematosus* – similar to rheumatoid arthritis.

c. *Psoriatic arthritis* – asymmetrical arthritis (**Fig. 15.73**).

d. *Gout* – initially peripheral monoarthritis with erythema (**Fig. 15.74**), later severe deformity with tophi (**Fig. 15.75**).

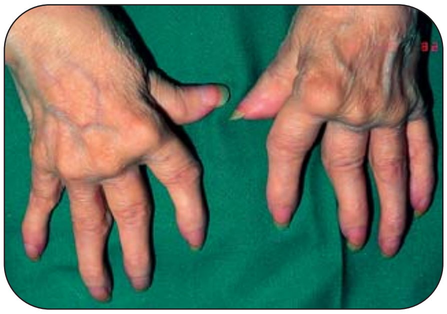

Fig. 15.72

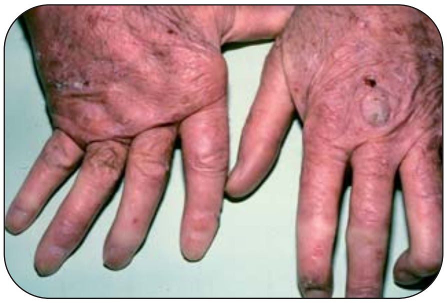

Fig. 15.73

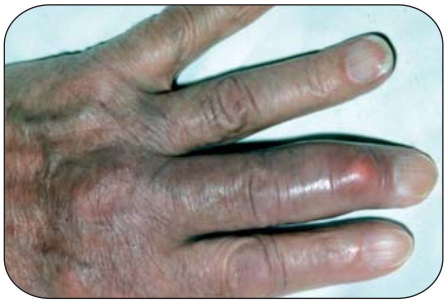

Fig. 15.74

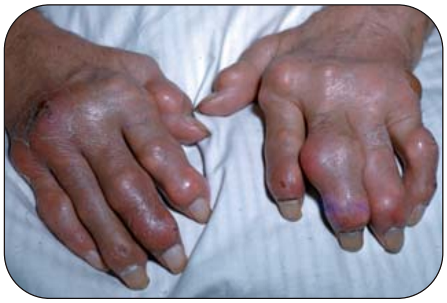

Fig. 15.75

e. *Juvenile idiopathic arthritis* – knees (**Fig. 15.76**).

f. *Reiter syndrome* – lower limbs (**Fig. 15.77**).

g. *Neuropathic (Charcot) arthropathy* – loss of joint sensation resulting in osteoarthritis with new bone formation and instability (**Fig. 15.78**).

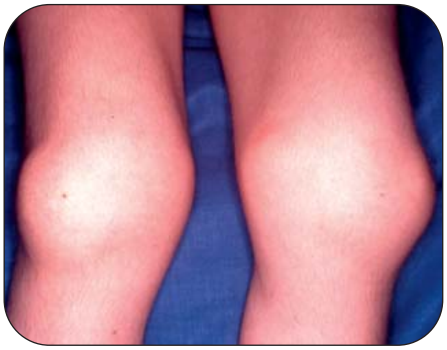

Fig. 15.76

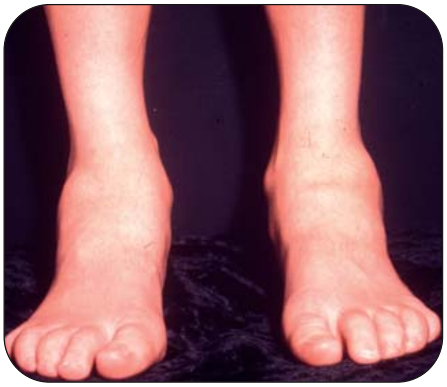

Fig. 15.77

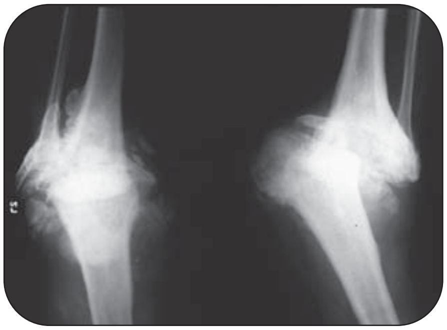

Fig. 15.78

SKIN

Erythema nodosum

Signs

- Tender, erythematous nodules on the knees and shins (**Fig. 15.79**), and occasionally on the thighs and forearms.

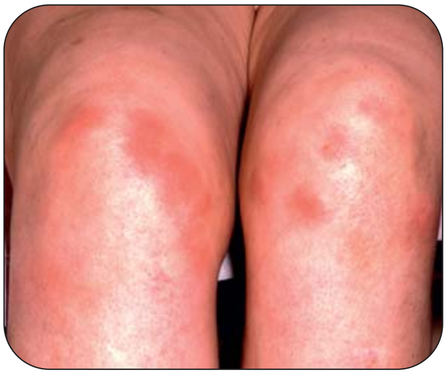

Fig. 15.79

Look for

- Acute sarcoidosis (Lofren syndrome).
- Tuberculosis.
- Crohn disease.
- Ulcerative colitis.
- Behçet disease.
- Lepromatous leprosy.
- Cat-scratch fever.

Vitiligo

Signs

- Well-defined, symmetrical areas of progressive depigmentation (**Fig. 15.80**).

Look for

- Vogt–Koyanagi–Harada syndrome.
- Sympathetic ophthalmitis.
- Myxoedema.
- Thyrotoxicosis.
- Pernicious anaemia.
- Autoimmune Addison disease.

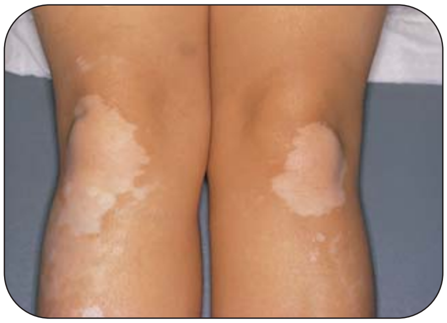

Fig. 15.80

Localised hypopigmented patches

Look for

- Lepromatous leprosy – numerous hypopigmented macules (**Fig. 15.81**).
- Tuberous sclerosis – sparse, subtle hypopigmented patches (ash leaf spots) on the trunk (**Fig. 15.82**), limbs and scalp.

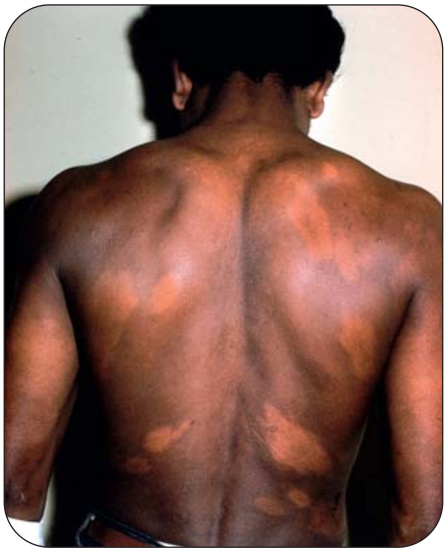

Fig. 15.81

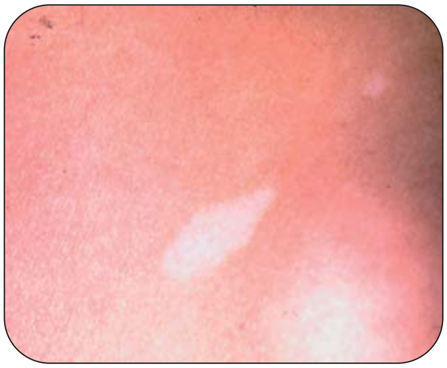

Fig. 15.82

Acanthosis nigricans

Signs

- Localised hyperpigmented patches in the axillae (**Fig. 15.83**) and groin.

Look for

- Carcinoma of stomach, breast or bronchus.

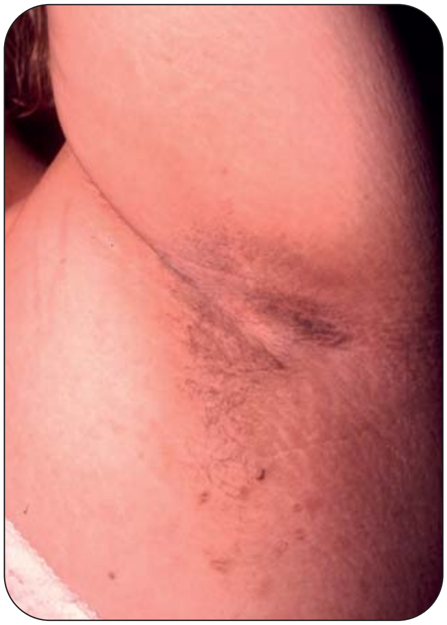

Fig. 15.83

Generalised hyperpigmentation

Look for

- Addison disease (**Fig. 15.84**).
- Haemochromatosis.
- Whipple disease.
- Cushing disease (see Fig. 15.18).
- Porphyria cutanea tarda.

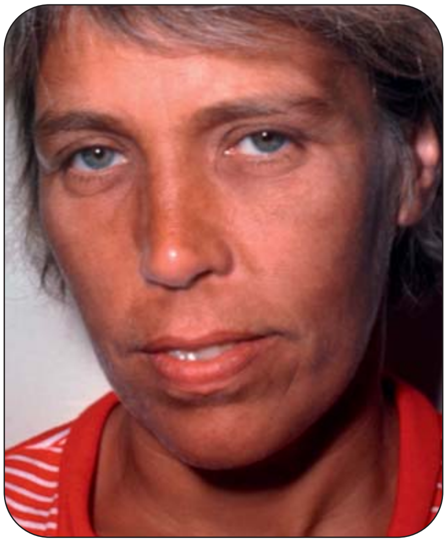

Fig. 15.84

Vesiculobullous dermatoses (Fig. 15.85)

Look for

The following which may be associated with cicatrising conjunctivitis:

- Mucous membrane pemphigoid.
- Stevens–Johnson syndrome (erythema multiforme major).
- Epidermolysis bullosa.
- Pemphigus vulgaris.
- Toxic epidermal necrolysis (Lyell disease, scalded skin syndrome).
- Pemphigoid.
- Linear IgA disease (bullous dermatosis).
- Dermatitis herpetiformis.

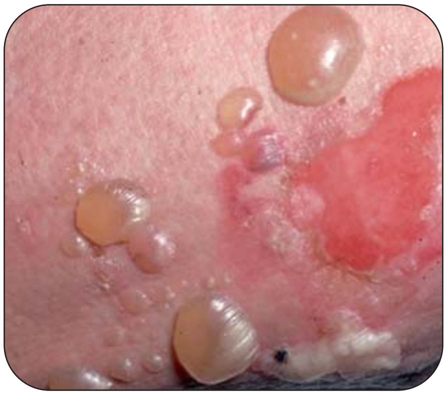

Fig. 15.85

Pyoderma gangrenosum

Signs

- Skin necrosis with irregular ulceration (**Fig. 15.86**).

Look for

- Wegener granulomatosis
- Ulcerative colitis.
- Crohn disease.
- Rheumatoid arthritis.

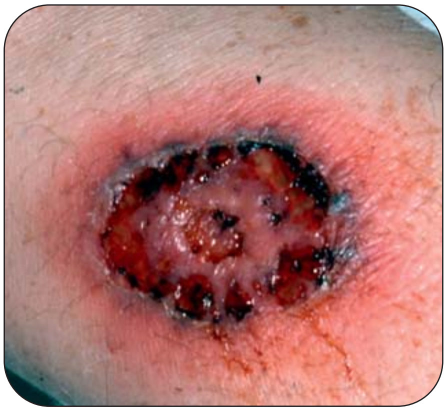

Fig. 15.86

Cutaneous bleeding

Signs

- Purpura, which is a small cutaneous extravasation of blood (**Fig. 15.87**).
- Ecchymosis or bruising is more extensive cutaneous bleeding (**Fig. 15.88**).

Look for

- Leukaemia.
- Polycythaemia rubra vera.
- Multiple myeloma.
- Hermansky–Pudlak syndrome.

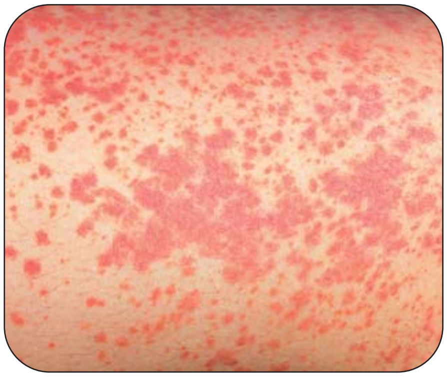

Fig. 15.87

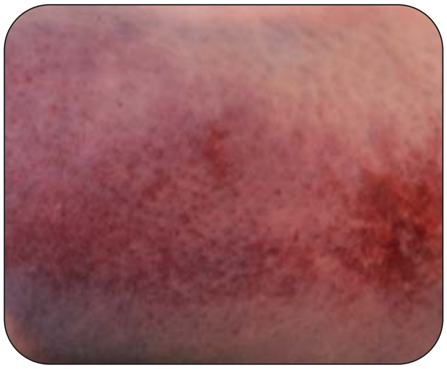

Fig. 15.88

Thin hyperelastic skin (Fig. 15.89)

Look for

- Ehlers–Danlos syndrome type 6.
- Pseudoxanthoma elasticum.

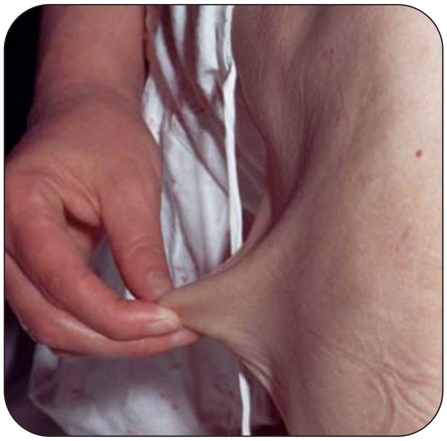

Fig. 15.89

Vasculitis

Signs

- Palpable purpura which may progress to necrosis or bullae formation typically affecting the lower parts of the legs (**Fig. 15.90**).

Look for

- Polyarteritis nodosa.
- Rheumatoid arthritis.
- Systemic lupus erythematosus.
- Churg–Strauss syndrome.

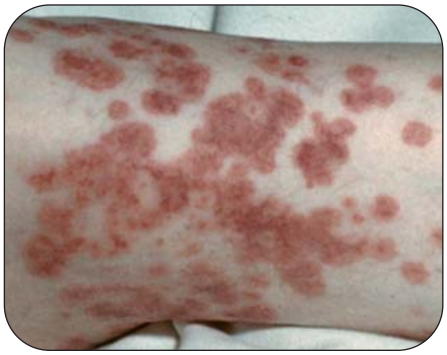

Fig. 15.90

Telangiectasia

Signs

- Dilated blood vessels which assume various shapes and blanch on pressure (**Fig. 15.91**).

Look for

- Ataxia telangiectasia (Louis-Barr syndrome).
- Fabry disease.
- Systemic lupus erythematosus.
- Systemic sclerosis.
- Dermatomyositis.

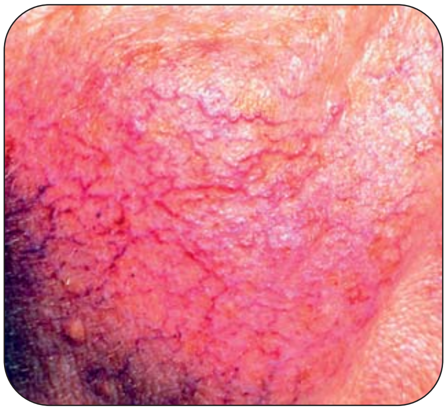

Fig. 15.91

Livedo reticularis

Signs

- Persistent, patchy, reddish-blue mottling of the legs which tends to be worse in cold weather (**Fig. 15.92**).

Look for

- Polyarteritis nodosa.
- Antiphospholipid antibody syndrome.

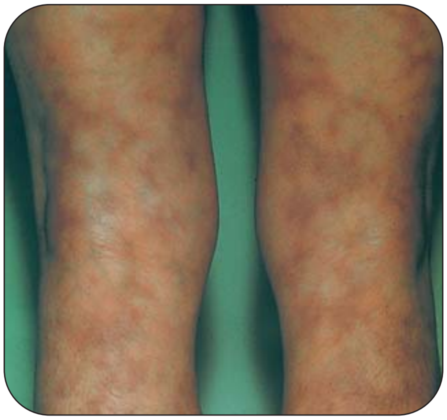

Fig. 15.92

Necrobiosis lipoidica

Signs

- Bilateral plaques with shiny atrophic centres on the shins (**Fig. 15.93**).

Look for

- Diabetes.
- AIDS.

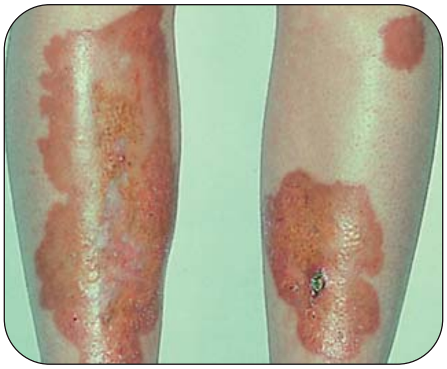

Fig. 15.93

Acquired ichthyosis

Signs

- Itchy dry and scaly skin which may be associated with hyperkeratosis (**Fig. 15.94**).

Look for

- Lymphoma.
- Refsum disease.

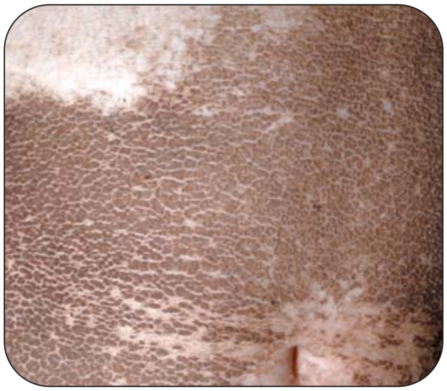

Fig. 15.94

Multiple tumours

Look for

- Xeroderma pigmentosum – multiple melanomas and carcinomas (**Fig. 15.95**).
- Kaposi sarcoma in AIDS (**Fig. 15.96**).
- NF1 – pedunculated neurofibromas (fibroma mollusca – **Fig. 15.97**).

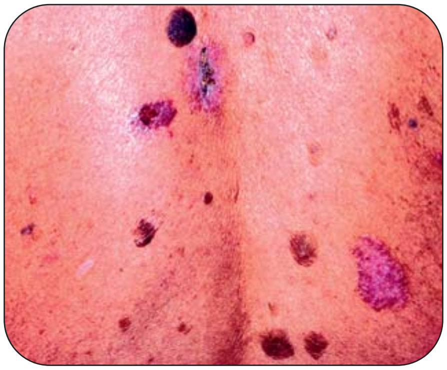

Fig. 15.95

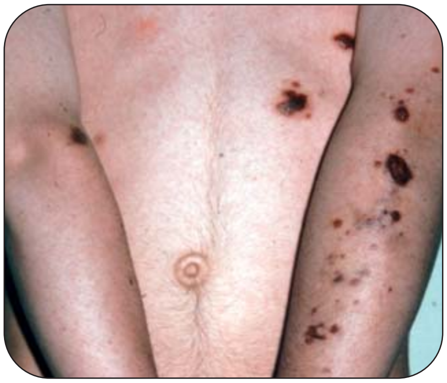

Fig. 15.96

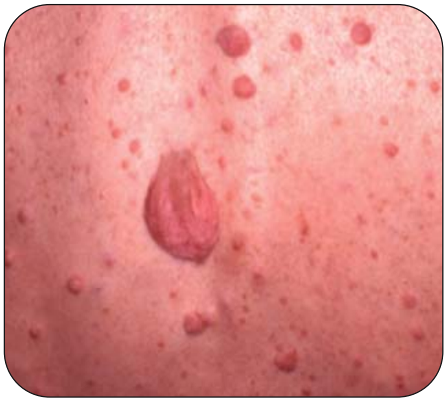

Fig. 15.97

GENITALIA

Urethritis (Fig. 15.98)

Look for

- Chlamydial infection.
- Gonorrhoea.
- Reiter syndrome.

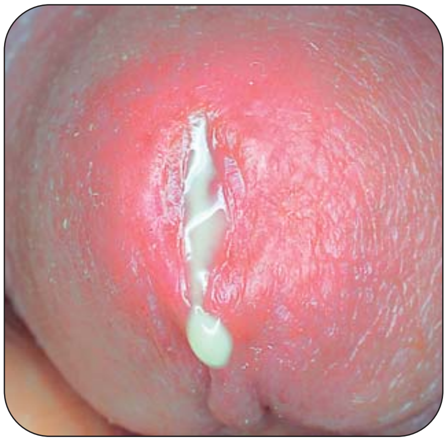

Fig. 15.98

Ulceration

Look for

- Behçet disease – recurrent ulceration of the scrotum or penis in males (**Fig. 15.99**) and the labia (**Fig. 15.100**) and vagina in females.
- Stevens–Johnson syndrome – bullae and erosions of the glans (**Fig. 15.101**).
- Reiter disease – painless erythematous erosion of the glans penis (circinate balanitis – **Fig. 15.102**).
- Acquired syphilis – painless ulcer (chancre – **Fig. 15.103**) with regional lymphadenopathy.
- Chancroid and lymphogranuloma venereum – genital ulceration and regional lymphadenopathy (**Fig. 15.104**).

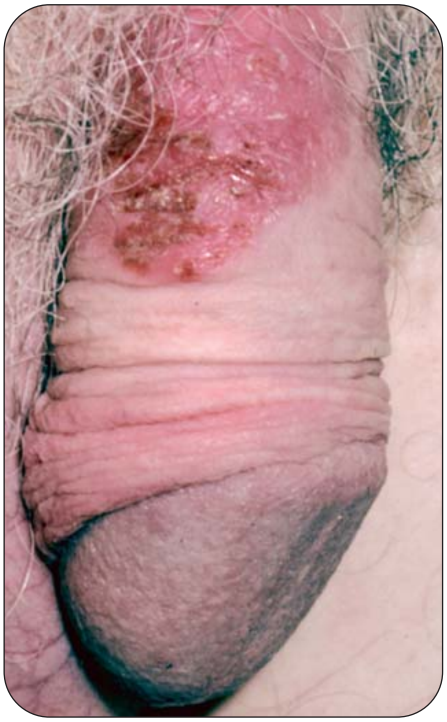
Fig. 15.99

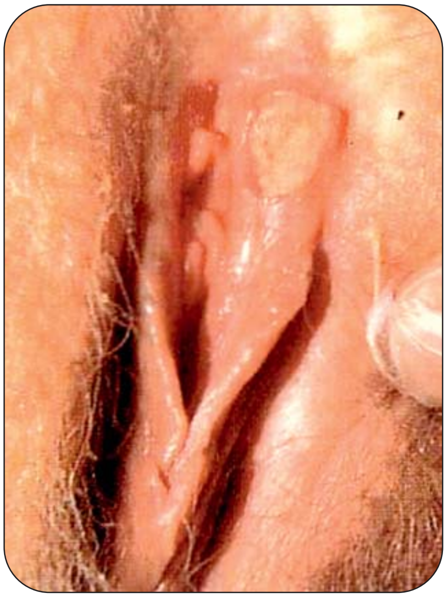
Fig. 15.100

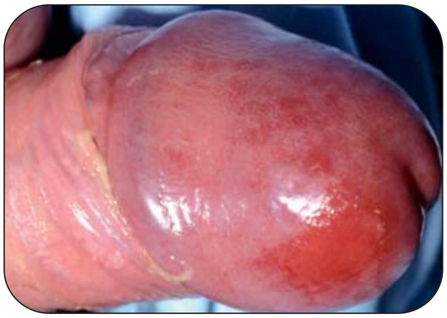
Fig. 15.101

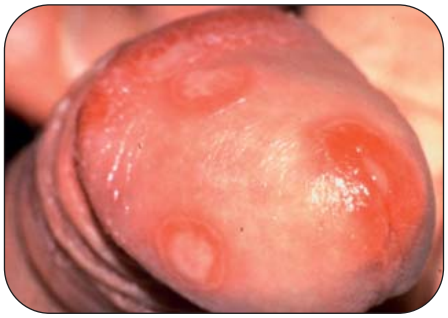
Fig. 15.102

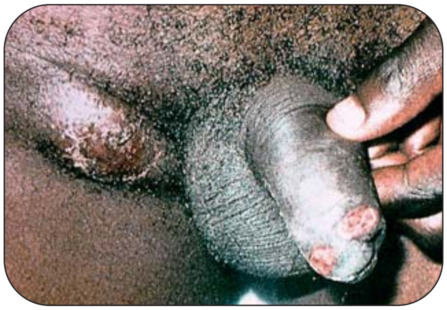
Fig. 15.104

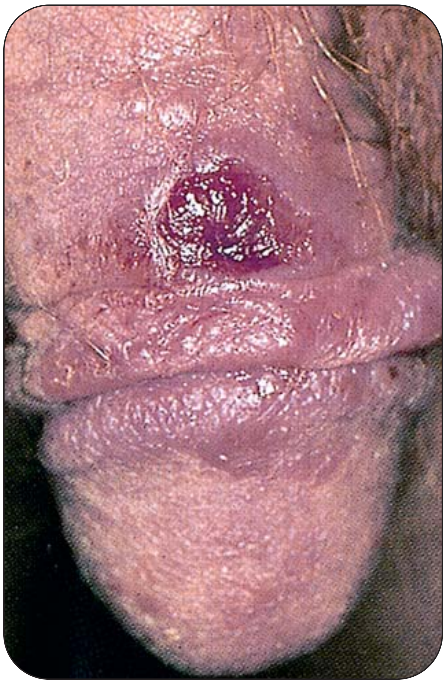
Fig. 15.103

Balanitis and phimosis

Signs

- Infection of the glans (**Fig. 15.105**).
- Constriction of the preputial orifice so that the prepuce cannot be retracted back over the glans (**Fig. 15.106**).

Look for

- Diabetes.

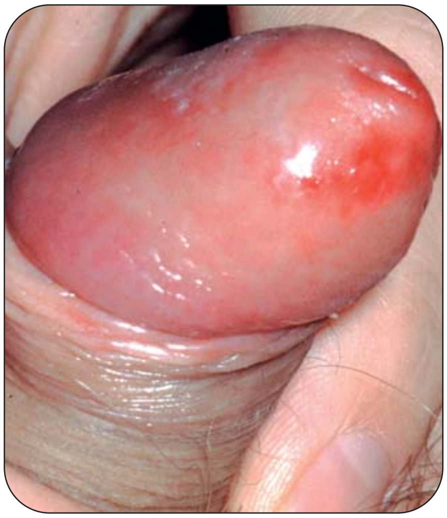

Fig. 15.105

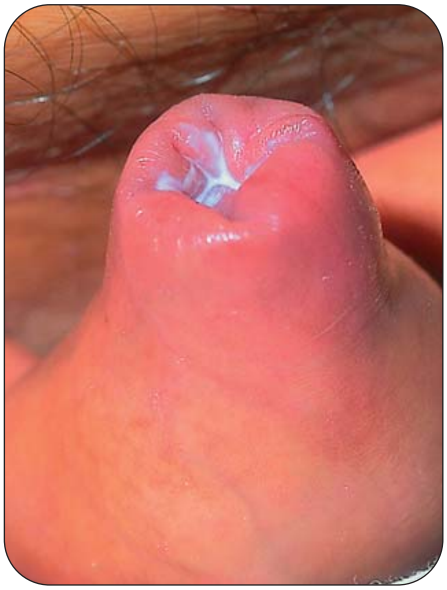

Fig. 15.106

Index

Note: Page numbers in **bold** refer to figures.

A

Abducens nucleus lesion, 376, **376**
Ablepharon, 37, **37**
Abscess, subperiosteal, 60
Acanthamoeba keratitis, 122, **122**
Acanthamoeba pseudodendrite, 118, **118**
Acanthosis nigricans, 414, **414**
Accelerated hypertension, **263**
Acne rosacea, 77, **77**, 385, **385**
Acquired cataract, 225–30, **225–30**
  anterior capsular, 225–6, **225–6**
  anterior subcapsular, 226, **226**
  cortical, 228–9, **228–9**
  nuclear, 230, **230**
  posterior subcapsular, 227–8, **227–8**
Acromegaly
  abnormal tongue, 403, **403**
  facial signs, 393, **393**
  large skull, 405, **405**
Actinic keratosis, 32, **32**
Active thyroid ophthalmopathy, 79, **79**
Acute angle-closure, 114, **114**
Acute stromal keratitis, 138, **138**
Adenoma sebaceum, 387, **387**
Adenoviral conjunctivitis, 66, **66**
Adie pupil, 198, **198,** 205
Adrenochrome deposits, 83, **83,** 123, **123**
Adult inclusion conjunctivitis, 69, **69**
Age-related macular degeneration
  dry, 317, **317**
  wet, 285, **285,** 289, **289**
Albinism, **181,** 181–2
  oculocutaneous, 394, **394**
Albinotic spots, congenital, 343, **343**
Alkaptonuria, 98, **98**
Allergic reactions, eyelid, 9, **9**
Allergic rhinoconjunctivitis, 65, **65**
Alphabet patterns, 366–7, **366–7**
Alport syndrome, 339, **339**
Amyloidosis
  deposits in familial, 243, **243**
  focal lesions, 88, **88**
  lacrimal gland enlargement, 59
  primary, 14, **14**
Angiitis, frosted branch, 301, **301**
Angioedema, 12, **12**
Angioid streaks, 272, **272,** 315, **315**
Angle closure, 151–62, 156–8, **156–8**
  lens-induced, 156, **156**
  post acute, 183, **183**
  primary, 159, **159**
  with pupil block, 156, **156**
  subacute, 155
  without pupil block, 157–8, **157–8**
Angle kappa
  negative, 361
  positive, 363, **363**
Angle recession, 155, **155**
Angular blepharitis, 8, **8**
Aniridia
  angle closure, 158, **158**
  cataracts in neonates, 221
  iris defect/absence, 186, **186**
Anisocoria, right physiological, 195, **195**
Ankylosing spondylitis, 167–8, **167–8,** 412, **412**
Anophthalmos with cyst, 51, **51**
Anterior capsule
  cataract, 225–6, **225–6**
  fibrosis and contraction (phimosis), 232, **232**
  pigmentation on, 226, **226**
Anterior chamber, 141–50
  abnormal depth, 143–5, **143–5**
    shallow, 143–5, **143–5**
    very deep, 143, **143**
  inflammatory signs, 145–7, **145–7**
  non-inflammatory cells, 148–50, **148–50**
  see also specific disorders
Anterior embryotoxon, 135, **135**
Anterior lenticonus, 214, **214**
Anterior orbital encephalocele, 51, **51**
Anterior uveitis, 145–6, **145–6,** 165–72, **165–72**
  acute, 165, **165**
  chronic, 165–6, **165–6**
  clinical features, 165–6, **165–6**
  herpes zoster and simplex, 185, **185**
  small pupil, 204, **204**
  systemic associations of, 167–72, **167–72**
  trabecular pigmentation, 155
  without systemic associations, 166–7, **166–7**
A pattern deviation, 366, **366**
Apert syndrome, 43, **43**
Aphakia, 162
Apocrine sweat gland hidrocystoma, 28, **28**
Aqueous cells, 145, **145**
Arachnodactyly, 406, **406**
Argyll Robertson pupils, 201, **201,** 204, **204**
Argyrosis, 83, **83,** 129, **129**
Arteriovenous communication, congenital, 295, **295**
Asteroid hyalosis, 242, **242**
Astrocytoma
  optic nerve head, 267, **267**
  retinal, 346, **346**
Ataxia-telangiectasia, 80, **80**
Atopic dermatitis, 9, **9**
Atopic eczema, 385, **385**
Atopic keratoconjunctivitis, **67,** 67–8
Atrophic maculopathy, 317–18, **317–18**
Autosomal dominant crystalline retinopathy, 322, **322**
Autosomal dominant neovascular inflammatory vitreoretinopathy, 245
Autosomal dominant vitreoretinochoroidopathy, 245
Avellino dystrophy, 131, **131**
Axenfeld-Reiger anomaly
  angle closure, 158, **158**
  iris atrophy/translucency, 182, **182**
  iris defect/absence, 187, **187**

B

Bacterial conjunctivitis, 65, **65**
  neonatal, 71
Bacterial keratitis, 121, **121**
Balanitis, 168, **168,** 420, **420**
Band keratopathy, 124, **124**
Basal cell carcinoma
  nodular, 33, **33**
  sclerosing, 36, **36**
  ulcerating, 35, **35**
Basal cell papilloma, 30, **30**
Basal laminar drusen, 337, **337**
Bassen-Kornzweig syndrome, 23–4
Behçet disease
  anterior uveitis, 169, **169**
  chorioretinitis, 303–4, **303–4**
  genitalia ulceration, 419, **419**
  mouth ulceration, 397, **397**
Benign concentric annular macular dystrophy, 319
Benign epithelial (racial) melanosis, 82, **82**
Benign flecked retina, 339, **339**
Benign hereditary intraepithelial dyskeratosis, 89, **89**
Benson disease, 242, **242**
Bergmeister papilla, 264, **264**
Best vitelliform macular dystrophy, 344, **344**
Biber-Haab-Dimmer lattice dystrophy, 130, **130**
Bielschowsky head tilt test, 369, **369,** 370, 371
Bietti crystalline dystrophy, 130, **130,** 322, **322**
Birdshot retinochoroidopathy, 308, **308**
Birth trauma, 109, **109**
Bleb, 145, **145**
  leaking, 144, **144**
Bleeding, cutaneous, 416, **416**

- Blepharitis
  - angular, 8, **8**
  - chronic, 8, 76, **76**
  - posterior, 7, **7**
  - seborrhoeic, 7, **7**
  - staphylococcal, 7, **7**
- Blepharochalasis, 13, **13**
- Blepharokeratoconjunctivitis, childhood, 8, **8**
- Blepharophimosis syndrome, 24, **24**
- Blepharospasm, essential, 391, **391**
- Blistering dermatoses, cicatrising conjunctivitis in, 72–4, **72–4**
- Bloch-Sulzberger syndrome, 207, **207,** 292
- Blond hair, fair complexion and, 394–5, **394–5**
- Blood dyscrasias, 148, 282, **282**
- Blow-out fracture
  - medial wall, 374, **374**
  - orbital floor, 374, **374**
- Blunt trauma
  - eyelid, 12, **12**
  - hyphaema, 148, **148**
  - iris, 188, **188**
  - ocular, 217
  - vitreous, 239, **239**
- Bowen disease, 36, **36**
- Bowman layer
  - degenerations in, 124–6, **124–6**
  - dystrophies, 126–7, **126–7**
- Brachydactyly, 406, **406**
- Brown syndrome, 364, **364**
- Brow ptosis, 25, **25**
- Bruits, 60
- Brushfield spots, 174, **174**
- Bullous pemphigoid, 73, **73**
- Bull's eye maculopathy, 319, **319**
- Buphthalmos, 105, **105**
- Buried optic disc drusen, 260–1, **260–1**

C

- Calcific emboli, 293, **293**
- Calibre changes, vascular, **297–8**
- Canaliculitis, chronic, 77, **77**
- Candida keratitis, 121, **121**
- Candidiasis, 302, **302**
- Canthaxanthin, 323, **323**
- Capillary haemangioma
  - eyelid, 31, **31**
  - fundus, 288, **288**
  - optic nerve head, 266, **266**
  - orbit, 47, **47**
  - proptosis, 60
  - retinal, 239, 295, **295,** 297, **298**
- Carotid-cavernous fistula, 13, **13,** 53, **53,** 79, **79**
- Cataract, 206, **206**
  - acquired *see* Acquired cataract
  - childhood *see* Childhood cataract
- Cat-scratch fever, 75, **75**
- Cavernous haemangioma
  - optic nerve head, 266, **266**
  - orbit, 54, **54**
  - retinal, 296, **296**
- Cavernous sinus thrombosis, 53
- Cellophane maculopathy, 320, **320**
- Cellulitis
  - orbital, 46, **46,** 53
  - preseptal, 46, **46**
- Central areolar choroidal dystrophy, 318, **318**
- Central cloudy dystrophy of François, 132, **132**
- Central pulverulent childhood cataract, 220, **220**
- Central retinal artery occlusion, acute, 345, **345**
- Central retinal vein occlusion, 258, **258**
- Central Schnyder (crystalline) dystrophy, 126, **126**
- Cerebrovascular accidents, 377
- Chalazion, 31, **31**
- Chancroid, 419, **419**
- Charcot arthropathy, 413, **413**
- Chemical neonatal conjunctivitis, 71, **71**
- Cherry-red spot, 345, **345**
- Childhood cataract, 218–25, **218–25**
  - anterior polar, 218, **218**
  - causes and associations
    - in neonates, 221–3, **221–3**
    - in older children, 224–5, **224–5**
  - central pulverulent, 220, **220**
  - coronary, 219, **219**
  - focal blue dot, 220, **220**
  - lamellar, 219, **219**
  - morphology, 218–20, **218–20**
  - posterior polar, 218, **218**
  - sutural, 220, **220**
- Children
  - blepharokeratoconjunctivitis, 8, **8**
  - cataract *see* Childhood cataract
  - glaucoma, 160–2, **160–2**
  - orbital disease in, 46–52, **46–52**
  - white pupil, 206–9, **206–9**
- Chlamydial keratitis, 136–7, **137**
- Chlamydial neonatal conjunctivitis, 71
- Chloroquine, 319, 332, **332**
- Cholesterol plaques, 293, **293**
- Chorioretinal atrophy
  - diffuse, 350–2, **350–2**
    - acquired, 351–2, **351–2**
    - hereditary, 350–1, **350–1**
  - glaucomatous, 271, **271**
  - idiopathic, 342, **342**
  - myopic, 272, **272**
  - pigmented paravenous, 334–5, **335**
  - progressive bifocal, 351, **351**
- Chorioretinal colobomas, 207, **207**
- Chorioretinal folds, 353, **353**
- Chorioretinitis
  - end-stage acquired syphilitic, 334, **334**
  - onchocercal, 352, **352**
  - with systemic associations, 301–6, **301–6**
  - without systemic associations, 306–9, **306–9**
- Chorioretinopathy, serpiginous, 352, **352**
- Choroid
  - circumscribed haemangioma, 348, **348**
  - coloboma, 343, **343**
  - detachment *see* Choroidal detachment
  - dystrophy *see* Choroidal dystrophy
  - focal elevated lesions, 347–9, **347–9**
  - granuloma, 349, **349**
  - haemorrhages, 279, **279**
  - inflammation *see* Choroiditis, multifocal, with panuveitis syndrome
  - melanoma, 268, **268,** 274, **274,** 347–8, **347–8**
  - metastatic carcinoma, 349, **349**
  - naevus, 341, **341**
  - neovascularisation, 272, **272,** 314–16, **314–16**
  - osteoma, 274, **274**
  - pneumocystosis, 305–6, **306**
- Choroidal detachment, 145, **145**
  - *vs.* retinal detachment, 332, **332**
- Choroidal dystrophy
  - central areolar, 318, **318**
  - generalised, 350, **350**
- Choroideremia, 350, **350**
  - female carriers of, 336, **336**
- Choroiditis, multifocal, with panuveitis syndrome, **306,** 306–7, 321, **321**
- Choroidopathy
  - punctate inner, 307, **307**
  - serpiginous, 274, **274,** 309, **309**
- Christmas tree cataract, 229, **229**
- Chronic myeloid leukaemia, 292
- Chronic toxocaral endophthalmitis, 209, **209**
- Chrysiasis, 129, **129**
- Cicatrical pemphigoid, 72, **72,** 73, **73,** 74
- Cicatrical retinopathy of prematurity, 208, **208**
- Cicatrising conjunctivitis in blistering dermatoses, 72–4, **72–4**
- Ciliary body melanoma
  - ectopia lentis, 217, **217**
  - with extraocular extension, 84, **84**
  - forward extension of, 174, **174**
- Ciliary body tumour, 158, **158**
- Coats disease, 209, **209,** 289, **289,** 294, **294**
- Cobb tufts, 148
- Cogan microcystic dystrophy, 112, **112**
- Cogan-Reese syndrome, 175, **175**
- Cogan syndrome, 134
- Collagen vascular disease, 283, **283**
- Collaterals, 264, **264**
- Collier sign of the midbrain, 18
- Colobomas
  - chorioretinal, 207, **207,** 343, **343**
  - eyelid, 38, **38**
  - iris, 187, **187**
  - lens, 213, **213**
  - optic nerve head, 253–4, **254,** 255, **255,** 270, **270**
- Comedones, 27, **27**
- Compressive thyroid optic neuropathy, 263
- Concomitant esotropia, 357–61, **357–61**
  - non-accommodative, 357–9, **357–9**
    - acute (late onset), 358

    - basic near, 357
    - consecutive, 359
    - convergence spasm, 359, **359**
    - cyclic, 359
    - distance, 358
    - early-onset (infantile), 357, **357**
    - microtropia, 358
    - sensory (secondary), 358–9, **359**
  - non-refractive accommodative, 360–1, **360–1**
  - pseudo-esotropia, 361, **361**
  - refractive accommodative, 360, **360**
- Concomitant exotropia, 361–3, **362–3**
  - consecutive, 362, **362**
  - constant (early onset), 361
  - hypertelorism, 362, **362**
  - intermittent (basic), 362, **362**
  - microexotropia, 362
  - pseudo-exotropia, 363, **363**
  - sensory (secondary), 362, **362**
- Cone dystrophy, 319, **319**
- Congenital anomalies/syndromes
  - anterior staphyloma, 110, **110**
  - cystic eyeball, 51, **51**
  - dacryostenosis, 72, **72**
  - eyelids, 37–8, **37–8**
  - fibrosis syndrome, 24, **24,** 365, **365**
  - glaucoma, 109, **109**
  - hereditary endothelial dystrophy, 111, **111,** 114
  - hereditary stromal dystrophy, 111
  - melanocytosis, **82,** 82–3
  - naevus, 16, **16**
  - ocular motility, 363–6, **363–6**
  - pit, 253, **253,** 312, **312**
  - *see also specific anomalies/syndromes*
- Congestive proptosis, 52–3, **52–3**
- Conjunctiva, 61–89
  - cystic lesions, 89, **89**
  - inflammation *see* Conjunctivitis
  - nodular lesions, 85–7, **85–7**
  - non-pigmented focal lesions, 85–9, **85–9**
  - pigmented lesions, 82–4, **82–4**
  - plaque-like lesions, 87–9, **87–9**
  - vascular dilatation and malformation, 79–81, **79–81**
  - *see also specific disorders*
- Conjunctival injection, 63, **63**
- Conjunctival intraepithelial hyperplasia, 86, **86**
- Conjunctivitis, 63–78, **63–78**
  - acute
    - follicular, 66, **66**
    - papillary, 65, **65**
  - adenoviral, 66, **66**
  - adult inclusion, 69, **69**
  - chronic
    - follicular, 69–70, **69–70**
    - papillary, 67–8, **67–8**
    - secondary, 76–8
    - toxic, 70, **70**
  - cicatrising, 72–4, **72–4**
  - diffuse eyelid swelling, 11, **11**
  - giant papillary, 68, **68**
  - gonococcal, 65, **65**
  - herpes simplex, 66, **66**
  - neonatal, 71–2, **71–2**
  - signs of inflammation, 63–4, **63–4**
  - simple bacterial, 65, **65**
  - in systemic disease, 74–6, **74–6**
- Contact dermatitis, 9, **9**
- Contralateral lid retraction, 25, **25**
- Convergence excess, **360,** 360–1
- Convergence spasm, 359, **359**
- Cornea, 101–40
  - abnormal shape, 106–8, **106–8**
  - abnormal size, 103–5, **103–5**
  - abrasion, 119, **119**
  - Bowman layer *see* Bowman layer
  - circumcorneal opacification, 134–5, **134–5**
  - dendritic and pseudodendritic ulceration, 117–18, **117–18**
  - Descemet membrane deposits, 127–30, **127–30**
  - epithelium
    - deposits, **123,** 123–4
    - iron lines, 123–4, **124**
    - large defects, 119–20, **119–20**
    - macroerosion, 120, **120**
    - microcysts, 112, **112**
    - oedema, 112, **112**
  - filaments, 116–17, **117**
  - infiltrates, 136–8, **136–8**
  - inflammation *see* Keratitis
  - large, 104–5, **104–5**
  - microbial ulcers, 121–2, **121–2**
  - oedema, 113–14, **113–14**
  - perinatal clouding, 109–11, **109–11**
  - peripheral lesions, 134–40, **134–40**
  - prominent nerves, 111, **111**
  - punctate lesions, 115–16, **115–16**
  - small, 103–4, **103–4**
  - stroma *see* Stroma
  - thinning and ulceration, 138–40, **138–40**
  - ulceration *see* Corneal ulceration
  - *see also specific disorders*
- Cornea farinata, 130, **130**
- Corneal arcus, 135, **135**
- Corneal ulceration
  - dendritic and pseudodendritic, 117–18, **117–18**
  - microbial, 121–2, **121–2**
  - and thinning, 138–40, **138–40**
- Cornea plana, 108, **108**
- Corneodysgenesis, 161
- Coronary childhood cataract, 219, **219**
- Cortical cataract, 228–9, **228–9**
- Cotton balls, 241, **241**
- Cotton-wool spots, 280–4, **280–4**
- Craniosynostoses, shallow orbits associated with, 42–4, **42–4**
- Criswick-Schepens syndrome, 245, **245,** 269, **269**
  - neovascularisation, 292, **292**
- Crohn disease, 138, 171
- Crouzon syndrome, 43, **43**
- Cryptophthalmos, 38, **38**
- Crystalline maculopathy
  - drug-induced, 323, **323**
  - hereditary, 321–2, **321–2**
  - miscellaneous, 324, **324**
- Cuneiform cataract, 228, **228**
- Cushing syndrome, 392, **392**
- Cutaneous bleeding, 416, **416**
- Cutaneous horn, 33, **33**
- Cuticular (basal laminar) drusen, 337, **337**
- Cyst/cystic lesions
  - anophthalmos with, 51, **51**
  - conjunctival, 89, **89**
  - deep dermoid, 49, 51, **51**
  - eyelids, 28–9, **28–9**
  - iris, 177–8, **177–8**
  - microphthalmos with, 51, **51**
  - orbital, 51–5, **51–5**
  - vitreous, 243–4, **243–4**
- Cysticercosis, 244, **244**
- Cystinosis, 127, **127,** 322
- Cyst of Moll, 28, **28**
- Cyst of Zeis, 29, **29**
- Cytomegalovirus retinitis, 289, **289,** 309, **309**

D

- Dacryoadenitis, 11, **11,** 57, **57**
- Dacryocystitis
  - chronic, 77, **77**
  - diffuse eyelid swelling, 11, **11**
- Dacryops, 59, **59**
- Dacryostenosis, congenital, 72, **72**
- Dalgleisch line, 124
- Deep dermoid cyst, 49, 51, **51**
- Degenerative retinoschisis, 331, **331**
- Dellen, 138–9, **139**
- Dendritic ulceration, 117, **117**
- Dental caries, 399, **399**
- Dentition, abnormal, 399–400, **399–400**
- Dermatitis
  - atopic, 9, **9**
  - contact, 9, **9**
- Dermatitis herpetiformis, 74, **74**
- Dermatochalasis, 25, **25**
- Dermatomyositis
  - cotton-wool spots, 283, **283**
  - eyelids, 16, **16**
  - signs, 387, **387**
- Dermatosis papulosa nigra, 28, **28**
- Dermoid cyst
  - deep, 49, 51, **51**
  - superficial, 59, **59**
- Descemet membrane, deposits in, 127–30, **127–30**
- Diabetic iridopathy, 184, **184**
- Diabetic papillopathy, 263, **263**
- Diabetic retinopathy, 280, **280,** 285, **285**
- Diffuse subretinal fibrosis, 321
- Direct carotid-cavernous fistula, 13, **13**
- Discharge in conjunctivitis, 64, **64**
- Disciform keratitis, 114, **114**

Disciform scar, 320, **320**
Distichiasis
  acquired, 5, **5**
  congenital, 5, **5**
Down-shoot
  in abduction, 373
  in adduction, 372
Down syndrome, 19, 224, **224**
Doyne honeycomb dystrophy, 337, **337**
Drug-induced disorders
  anterior uveitis, 167
  cataract, 227
  chorioretinal atrophy, 351
  crystalline maculopathy, 323, **323**
Drusen, 287, **287,** 297, **298**
  buried optic disc, 260–1, **260–1**
  choroidal neovascularisation, 315, **315**
  crystalline maculopathy, 324, **324**
  cuticular (basal laminar), 337, **337**
  familial, 337, **337**
  hard, 336, **336**
  peripheral, 327, **327**
  soft, 337, **337**
Dry age-related macular degeneration, 317, **317**
Duane retraction syndrome, 19, **19,** 45
  type 1, 363, **363**
  type 2, 363
  type 3, 364, **364**
Dyskeratosis, benign hereditary intraepithelial, 89, **89**
Dystopia, 42, **42**

E

Eales disease, 292, **292**
Eccrine sweat gland hidrocystoma, 28, **28**
Ectopia lentis, 162, 215–17, **215–17**
  acquired, 217, **217**
  developmental, 215–17, **215–17**
  hereditary causes without systemic manifestations, 215, **215**
  ocular associations, 215, **215**
Ectopia lentis et pupillae, 205, **205**
Ectropion uveae, 179, **179**
Eczema, atopic, 385, **385**
Egg-yolk focal lesions, 344, **344**
Ehlers-Danlos syndrome, 99, **99,** 217
Electric shock, cataract, 227, **227**
Elschnig pearls, 232, **232**
Emboli, retinal, 292–3, **292–3**
Embryonal sarcoma, 49, **49**
Embryotoxon
  anterior, 135, **135**
  posterior, 135, **135**
Emulsified silicone oil, 150, **150**
Encephalocele
  anterior orbital, 51, **51**
  posterior orbital, 52, **52**
Endophthalmitis, 146–7, **146–7**
  acute postoperative, 114, **114,** 146, **146**
  chronic
    postoperative, 147, **147**
    toxocaral, 209, **209**
  delayed-onset, 147, **147**
  endogenous infectious, 147
  infectious, 147, 238
Endophytic retinoblastoma, 240, **240**
Endothelial dystrophy
  congenital hereditary, 111, **111,** 114
  Fuchs, 113, **113**
Endothelium
  dystrophy *see* Endothelial dystrophy
  iatrogenic damage, 113, **113**
Enhanced S-cone syndrome, 335, **335**
Enophthalmos, 44–5, **44–5**
Epiblepharon, 6, **6**
Epibulbar dermoid, 86, **86**
Epicanthic folds, 361, **361**
Epidermal inclusion cyst, 29, **29**
Epidermolysis bullosa, 73, **73**
Episclera *see* Sclera/episclera
Episcleritis
  diffuse (simple), 93, **93**
  nodular, 94, **94**
Epitheliopathy, acute posterior multifocal placoid pigment, 308, **308**
Epithelium
  corneal
    deposits, **123,** 123–4
    iron lines, 123–4, **124**
    large defects, 119–20, **119–20**
    macroerosion, 120, **120**
    microcysts, 112, **112**
    oedema, 112, **112**
  iris, cystic lesions, 177, **177**
Erosive vitreoretinopathy, 245
Erysipelas, 10, **10**
Erythema nodosum, 413, **413**
Erythrocytes, 238–9, **238–9**
Esotropia, concomitant *see* Concomitant esotropia
Essential blepharospasm, 391, **391**
Ethmoidal/frontal carcinoma, 56
Ewing sarcoma, metastatic, 50
Excyclotorsion, 371
Exfoliation, true, 225, **225**
Exotropia, concomitant *see* Concomitant exotropia
Exposure keratopathy, 120, **120,** 137
External hordeolum, 31, **31**
Exudative retinal detachment, 331, **331**
Eyelashes *see* Lashes
Eyelid ptosis, 19–26, **19–26**
  aponeurotic, 22, **22**
  eyelash causing, 5, **5**
  mechanical, 22, **22**
  myogenic
    adult, 20–2, **20–2**
    childhood, 24–5, **24–5**
  neurogenic
    adult, 19–20, **19–20**
    childhood, 23–4, **23–4**
  simple congenital, 24, **24**
  see also Pseudo-ptosis
Eyelid retraction, 17–19, **17–19**
  contralateral, 25, **25**
Eyelids, 1–38
  blunt trauma, 12, **12**
  congenital inversion, 38, **38**
  cystic lesions, 28–9, **28–9**
  diffuse swelling, 11–14, **11–14**
  discoloration, 15–16, **15–16**
  infections, 10–11, **10–11**
  inflammatory margin disease, 7–9, **7–9**
  lack of globe support, 26, **26**
  lashes *see* Lashes
  major congenital anomalies, 37–8, **37–8**
  malignant tumours, 57
  multiple papules and vesicles, 26–8, **26–8**
  nodular lesions, 31–5, **31–5**
    benign, 31–3, **31–3**
    malignant, 33–5, **33–5**
  papillomatous lesions, 30, **30**
  plaque-like lesions, 36–7, **36–7**
  ptosis *see* Eyelid ptosis
  retraction *see* Eyelid retraction
  superficial inflammation, 9–11, **9–11**
  ulcerating tumours, 35–6, **35–6**
  see also specific conditions
Eye popping reflex, transient, 19

F

Fabry disease, 81, **81,** 224, **224**
Face
  fair complexion and blond hair, 394–5, **394–5**
  hemispasm, 392, **392**
  hirsutism, 392–4, **392–4**
  lack of expression, 389–91, **389–91**
  pigmentary changes, 388, **388**
  rash, 385–7, **385–7**
  saddle-shaped nose, 395–6, **395–6**
  spasm, 391–2, **391–2**
Facial nerve palsy, 18, **18**
Fair complexion and blond hair, 394–5, **394–5**
Familial drusen, 337, **337**
Familial exudative vitreoretinopathy, 245, **245,** 269, **269**
  neovascularisation, 292, **292**
Familial lecithin-cholesterol-acyltransferase deficiency, 128, **128**
Fenestrated sheen macular dystrophy, 319
Ferry line, 123
Fibrinoplatelet emboli, 292, **292**
Fibrinous exudate, 146, **146**
Fibrosarcoma, 50
Fibrosis syndrome, congenital, 24, **24**
Fibrotic fundus lesions, 320–1, **320–1**
Fibrous dysplasia, 49
Fibrous plaques, 226, **226**
Filamentous keratitis, 121–2, **122**
Filtration, excessive, 145, **145**
Filtration surgery, shallow chamber following, 144–5, **144–5**
Fingers
  clubbing, 407, **407**
  long (arachnodactyly), 406, **406**
  loss of, 408, **408**
  short stubby (brachydactyly), 406, **406**

Fish eye disease, 129
Fistula
  carotid-cavernous, 53, **53,** 79, **79**
  direct carotid-cavernous, 13, **13**
Flame-shaped haemorrhages, **277,** 277–8
Flare, aqueous, 145, **145**
Fleck dystrophy, 132
Fleischer ring, 124
Floaters, 241, **241**
Flocculi, iris, 177, **177**
Floppy eyelid syndrome, 77–8, **78**
Flower-shaped cataract, 229, **229**
Fluoroquinolones, 123, **123**
Focal blue dot, 220, **220**
Follicles
  acute conjunctivitis, 66, **66**
  chronic conjunctivitis, 69–70
  reaction in conjunctivitis, 63, **63**
Foreign bodies/material, vitreous, 244, **244**
Foster Kennedy syndrome, 258, **258**
Fourth nerve palsy, 368–9, **368–9**
Freckle
  conjunctival, 82, **82**
  iris, 174, **174**
Frosted branch angiitis, 301, **301**
Frozen orbit, 374–5, **375**
Fuchs endothelial dystrophy, 113, **113**
Fuchs syndrome
  anterior uveitis, 166, **166**
  heterochromia iridis, 191, **191**
  iris atrophy and translucency, 181, **181**
  vitritis, 238
Fundus, 275–354
  calibre changes, 297–8, **297–8**
  elevated focal lesions, 346–9, **346–9**
  extensive white lesions, 207, **207**
  fibrotic lesions, 320–1, **320–1**
  flat focal lesions, 340–5, **340–5**
    black, 340–2, **340–2**
    cherry-red spot, 345, **345**
    egg-yolk, 344, **344**
    pale, 342–3, **342–3**
  inflammatory lesions, 299–310, **299–310**
  multiple yellow-white spots and flecks, 336–40, **336–40**
  scattered pigmentary changes, 332–6, **332–6**
  vascular lesions, 277–98, **277–98**
  see also specific disorders
Fundus albipunctatus, 338, **338**
Fundus flavimaculatus, 338, **338**
Furrow degeneration, 138, **138**

G

Galactokinase deficiency, childhood cataracts, 224
Galactosaemia, 222–3, **222–3**
Gaze palsies, 376–7, **376–7**
Gelatinous drop-like dystrophy, 127, **127**
Genitalia, 418–20, **418–20**
Geographic ulcer, herpes simplex, 119, **119**
Giant papillary conjunctivitis, **68**
Gland of Zeis carcinoma, ulcerating, 36, **36**
Glaucoma
  childhood, 160–2, **160–2**
  congenital, 109, **109**
  malignant, 145, **145**
  neovascular, 159, **159**
  phacolytic, 149, **149**
  phacomorphic, 156, 159, **159**
  primary, 160–1, **160–1**
  secondary, 162
Glaucomatocyclitic crisis, 159, **159**
Glaucomatous chorioretinal atrophy, 271, **271**
Glaucomatous cupping, 256, **256**
Glaukomflecken, 226, **226**
Glioma, optic nerve, 48, **48**
Gonococcal conjunctivitis, 65, **65**
  neonatal, 71, **71**
Gout, 412, **412**
Granular dystrophy
  type 1, 131, **131**
  type 2, 131, **131**
Granulocytic granuloma, 50, **50**
Granuloma
  choroidal, 349, **349**
  granulocytic, 50, **50**
  pyogenic, 85–6, **86**
  sarcoid, 268, **268**
  toxocara *see* Toxocara granuloma
Guttering, asymptomatic, 140, **140**
Gyrate atrophy, 324, **324,** 350, **350**

H

Haemangioblastoma *see* Capillary haemangioma
Haemangioma
  capillary *see* Capillary haemangioma
  cavernous *see* Cavernous haemangioma
  circumscribed choroidal, 348, **348**
  retinal racemose, 296, **296**
Haematoma, subperiosteal, 59
Haemorrhages
  choroidal, 279, **279**
  in conjunctivitis, 64, **64**
  flame-shaped, **277,** 277–8
  fundus, 277–9, **277–9**
  intraretinal, 278, **278**
  optic nerve head, 265, **265**
  preretinal (subhyaloid), 277, **277**
  subretinal, 278–9, **279**
  sub-RPE, 279, **279**
  subungual splinter, 411, **411**
Haemorrhagic lymphangiectasia, 80, **80**
Haemorrhagic telangiectasia, hereditary, 81, **81**
Hallermann-Streiff-François syndrome, 223, **223**
Hamartoma
  combined, of the retina and RPE, 267, **267,** 270, **270**
  retinal pigment epithelium, 341, **341**
Hands, 406–9, **406–9**
  finger clubbing, 407, **407**
  hyperextensibility of joints, 407, **407**
  long fingers (arachnodactyly), 406, **406**
  loss of digits, 408, **408**
  polydactyly, 408, **408**
  prayer sign, 407, **407**
  Raynaud phenomenon, 408, **408**
  sclerodactyly, 408, **408**
  short stubby fingers (brachydactyly), 406, **406**
  syndactyly, 409, **409**
  wasting of small muscles, 406, **406**
Harada disease, 313
Hard drusen, 336, **336**
Hard exudates
  retinal, 284–7, **284–7**
  subretinal, 289, **289**
Hemispasm, facial, 392, **392**
Hereditary haemorrhagic telangiectasia, 81, **81**
Herpes simplex
  anterior uveitis, 185, **185**
  conjunctivitis, 66, **66**
  dendritic ulcer, 117, **117**
  eyelids, 26, **26**
  geographic ulcer, 119, **119**
  hyphaema, 148
Herpes zoster
  anterior uveitis, 185, **185**
  hyphaema, 148
  pseudodendrite, 118, **118**
Herpes zoster ophthalmicus, 10, **10,** 96, **96**
Heterochromia iridis, 189–91, **189–91**
  acquired, 191–2, **191–2**
  congenital, 189–90, **189–90**
Hidrocystoma
  apocrine sweat gland, 28, **28**
  eccrine sweat gland, 28, **28**
High myopia, 105, **105,** 253, **253,** 351, **351**
Hirschsprung disease, 190, **190**
Hirsutism, facial, 392–4, **392–4**
Histiocytosis
  Langerhans-cell, 49, **49**
  sinus, 49
Histoplasmosis syndrome, presumed ocular, 273, **273,** 307, **307**
HIV microangiopathy, 281, **281**
HLA-B27 associated anterior uveitis, 167
Hollenhurst plaques, 293, **293**
Homocystinuria
  ectopia lentis, 216, **216**
  facial signs, 395
  high-arched palate, 401, **401**
  long fingers, 406, **406**
Honeycomb retinal degeneration, 327, **327**
Horizontal gaze palsy, 376, **376**
Horner syndrome
  eyelids, 20, **20,** 23, **23**
  heterochromia iridis, 190, **190**
  right, 199, **199**
  small pupils, 205
Hudson-Stähli line, 123
Human immunodeficiency virus microangiopathy, 281, **281**
Hurler syndrome, 128
Hutchinson freckle, 15, **15**
Hyaline plaque, scleral, 98, **98**

Hyaloid artery, persistent, 243, **243**
Hydrocephalus, infantile, 18, **18**
Hydrops, 106, **106,** 113, **113**
Hyperelastic skin, thin, 416, **416**
Hyperlysinaemia, ectopia lentis, 217
Hypermetropia, 251, **251**
Hyperpigmentation
  generalised, 415, **415**
  trabecular, 153–5, **153–5**
Hyperplasia
  conjunctival intraepithelial, 86, **86**
  reactive lymphoid, 88, **88**
  reactive pseudo-epitheliomatous, 88, **88**
Hypertelorism, **41,** 41–2, 362, **362**
Hypertension
  calibre changes, 297, **297**
  malignant, **263**
  severe, 287, **287**
Hypertensive retinopathy, severe, 281, **281**
Hyperviscosity, 297, **298**
Hyphaema, 148, **148**
Hypoaccommodative convergence excess, 361
Hypopigmentation, 414, **414**
Hypoplastic disc, 252, **252**
Hypoproteinaemias, **128,** 128–9
Hypopyon, 146, **146**

I

Ichthyosis, acquired, 418, **418**
Idiopathic central serous retinopathy, 312, **312**
Idiopathic orbital inflammatory syndrome, 46–7, **47,** 53
Immunoprotein corneal deposits, 128, **128**
Impetigo, 10, **10**
Implantation cysts, 178, **178**
Inclusion conjunctivitis, 69, **69**
Inclusion cyst, conjunctival, 89, **89**
Incontinentia pigmenti, 207, **207,** 292
Incyclotorsion, 371
Infantile hydrocephalus, 18, **18**
Infectious crystalline keratopathy, 125–6, **126**
Infectious endophthalmitis, 238
Inferior oblique
  overaction, 372, **372**
  palsy, 371, **371**
Inferior rectus palsy, 370, **370**
Inflammation
  anterior chamber, 145–7, **145–7**
  choroid, **306,** 306–7, 321, **321**
  chronic intraocular, 227, **227**
  conjunctival *see* Conjunctivitis
  cornea *see* Keratitis
  diffuse eyelid swelling, 11–12, **11–12**
  episclera *see* Episcleritis
  fundus, 299–310, **299–310**
  intraocular, 162
  iris nodules, 175, **175**
  juxtapapillary lesions, 273–4, **273–4**
  lens plaques, 232–3, **233**
  lens precipitates, 233, **233**
  necrotising scleritis with, 95, **95**
  orbit, 46–7, **46–7**
  retina *see* Retinitis
  sclera *see* Scleritis
  superficial eyelid, 9–11, **9–11**
  vitreous opacities, 241–2, **241–2**
Inflammatory bowel disease, 138, 171, **171**
Inflammatory lid margin disease, 7–9, **7–9**
Inflammatory orbital disease, 12, **12**
Innocuous floaters, 241, **241**
Interferon, 283, **283**
Intermediate uveitis, 237, **237**
Interpupillary distance, narrow, 361
Interstitial keratitis, 133–4, **133–4**
Intracranial pressure, raised, 261
Intraepidermal carcinoma in situ, 36, **36**
Intraepithelial dyskeratosis, benign hereditary, 89, **89**
Intraocular inflammation, chronic, 227, **227**
Intraocular pressure, 151–62
  acute elevation of, 159–60, **159–60**
Intraocular tumours, 162, **162**
Intraretinal haemorrhage, 278, **278**
Intrauterine infections
  cataracts in neonates, 222, **222**
  rubella, 109
Inverted follicular keratosis, 30, **30**
Inverted Y pattern deviation, 367
Ipsilateral hypotropia, 26, **26**
Iridescent particles, vitreous, 242–3, **242–3**
Iridocorneal endothelial syndrome, 157, 187–8, **188**
Iridocyclitis, 237, **237**
Iridodysgenesis, 161, **161**
Iridoplegia, right traumatic, 203, **203**
Iridoschisis, 183, **183**
Iris, 163–91
  anterior uveitis *see* Anterior uveitis
  atrophy and translucency, 181–5, **181–5**
  coloboma, 187, **187**
  cystic lesions, 177–8, **177–8**
  ectropion uveae, 179, **179**
  flocculi, 177, **177**
  full-thickness defect or absence, 186–8, **186–8**
  melanoma, 155, **155**
  rubeosis iridis *see* Rubeosis iridis
  spots and nodules, 172–7, **172–7**
    multiple, 174–7, **174–7**
    solitary, 172–4, **172–4**
  tumours, 148, **148,** 179, **179,** 191, **191**
  see also specific disorders
Iron deficiency anaemia, 403, **403**
Iron lines, epithelial, 123–4, **124**
Isolated trabeculodysgenesis, 160, **160**

J

Juvenile idiopathic arthritis, 171, **171,** 413, **413**
Juvenile ossifying fibroma, 49
Juxtapapillary lesions, 270–4, **270–4**
  congenital, 270–1, **270–1**
  degenerative, 271–2, **271–2**
  inflammatory, 273–4, **273–4**
  neoplastic, 274, **274**

K

Kaposi sarcoma, 34, **34,** 418, **418**
Kawasaki disease, 75, **75**
Kayser-Fleischer ring, 127, **127,** 135
Keratitis
  acanthamoeba, 122, **122**
  acute stromal, 138, **138**
  bacterial, 121, **121**
  candida, 121, **121**
  chlamydial, 136–7, **137**
  disciform, 114, **114**
  filamentous, 121–2, **122**
  marginal, 136, **136**
  nummular, 116, **116**
  peripheral ulcerative, 140, **140**
  punctate epithelial, **115,** 115–16
  rosacea, 136, **136**
  sclerosing, 137–8, **138**
  superficial punctate, 115, **115**
Keratoacanthoma, 32, **32**
Keratoconjunctivitis
  atopic, **67,** 67–8
  superior limbic, 68, **68,** 117, **117**
  vernal, **67,** 67–8
Keratoconjunctivitis sicca (KCS), 76, **76,** 116–17, **117**
Keratoconus, 106, **106**
  posterior, 107–8, **108**
Keratoglobus, 107, **107**
Keratolysis, 140, **140**
Keratopathy
  band, 124, **124**
  exposure, 120, **120,** 137
  infectious crystalline, 125–6, **126**
  lipid, 125, **125**
  neurotrophic, 119, **119**
  vortex, 123, **123**
Keratosis
  actinic (solar, senile), 32, **32**
  inverted follicular, 30, **30**
  seborrhoeic, 30, **30**
Koilonychia, 410, **410**

L

Lacquer cracks, 315, **315**
Lacrimal gland
  carcinoma, 58, **58**
  enlargement, 57–9, **57–9**
    bilateral, 59, **59**
    unilateral, 57–9, **57–9**
  lesions, 57
Lambda pattern deviation, 367
Lamellar childhood cataract, 219, **219**
Langerhans-cell histiocytosis, 49, **49,** 405, **405**

Laser iridotomy, 188, **188**
Lashes
  disorders, 3–6
  metaplastic, 5, **5**
  misdirection, 4–6, **4–6**
  ptosis, 5, **5,** 26, **26**
Lattice dystrophy
  type 1, 130, **130**
  type 2, 130–1, **131**
  type 3, 131, **131**
Lattice retinal degeneration, 328, **328**
Leber congenital amaurosis, 319, **319**
Lecithin-cholesterol-acyltransferase deficiency, familial, 128, **128**
Leiomyoma, iris, 173, **173**
Lens, 211–34
  cataract *see* Cataract
  dislocated, 143, **143**
  ectopia lentis *see* Ectopia lentis
  fragments, 244, **244**
  non-opaque defects, 230–1, **230–1**
  opacities in pseudophakia, 231–4, **231–4**
  shape abnormalities, 213–15, **213–15**
  total dislocation, 156
  total opacification, 234, **234**
  see also specific disorders
Lens-induced angle closure, 156, **156**
Lens-induced anterior uveitis, 167
Lenticonus
  anterior, 214, **214**
  posterior, **214,** 214–15
Lentiglobus, 214, **214**
Lentigo maligna, 15, **15**
Leopard spots, 333, **333**
Lepromatous iritis, 185, **185**
Lepromatous miosis, 204, **204**
Leprosy, 396, **396**
Leucocytes, 237–8, **237–8**
Leukaemia
  chronic myeloid, 292
  orbital disease, 57
Leukaemia cells, 149, **149**
Leukocoria, 206–9, **206–9**
  acquired, 208–9, **208–9**
  congenital, 206–7, **206–7**
Leukonychia, 410, **410**
Lipid keratopathy, 125, **125**
Lipodermoid, 87, **87**
Lisch nodules, 174–5, **175**
Livedo reticularis, 417, **417**
Louis-Bar syndrome, 80, **80**
Lowe oculocerebrorenal syndrome, 221–2, **222**
Lowe syndrome, 161
Lupus pernio *see* Sarcoidosis
Lyell disease, 73, **73**
Lyme disease, **134**
Lymphadenopathy, preauricular, 64, **64**
Lymphangioma, 47–8, **48,** 60
Lymphogranuloma venereum, 134, **134,** 419, **419**
Lymphoid tumours, 55, **55**
Lymphomas, 59
  non-Hodgkin, 88, **88**
  primary intraocular, 240, **240**

M

Macroaneurysm, 239, **239**
Macular dystrophy, 132, **132**
  adult vitelliform, 344, **344**
  benign concentric annular, 319
  Best vitelliform, 344, **344**
  fenestrated sheen, 319
  North Carolina, 340, **340**
  Stargardt, 317–18, **318,** 319
Macular hole, 325–6, **325–6**
  idiopathic (age-related), 325, **325**
  microhole, 325, **325**
  myopic, 326, **326**
  trauma, 326, **326**
Macular lesions, 310–26, **310–26**
  see also specific lesions
Macular oedema, 310–11, **310–11**
Macular pseudo-hole, 326, **326**
Maculopathy
  atrophic, 317–18, **317–18**
  bull's eye, 319, **319**
  cellophane, 320, **320**
  crystalline *see* Crystalline maculopathy
  myopic, 317, **317**
  stellate, 287–9, **287–9**
Madarosis, 3, **3**
Malattia levantinese, 338, **338**
Malignant glaucoma, 145, **145**
Malignant hypertension, 263, **263**
Mammillations, iris, 175, **175**
Mannosidosis, 225
Marcus Gunn jaw-winking syndrome, 17–18, **18,** 23, **23**
Marfan syndrome
  ectopia lentis, 215–16, **215–16**
  high-arched palate, 400, **400**
  long fingers, 406, **406**
Marginal keratitis, 136, **136**
Maroteaux-Lamy syndrome, 128
Masquerade syndrome, 78, **78**
Maxillary carcinoma, 56, **56**
Medial longitudinal fasciculus lesion, 376, **376**
Medial wall blow-out fracture, 374, **374**
Meesman dystrophy, 112, **112**
Megalocornea, 104, **104,** 143, **143,** 154, **154**
Megalopapilla, 254–5, **255,** 256
Meibomian cyst, 31, **31**
Meibomian gland carcinoma
  nodular, 34, **34**
  spreading, 37, **37**
Melanocytic proliferation, bilateral diffuse uveal, 342, **342**
Melanocytoma, 84, **84**
  iris, 173, **173**
  optic nerve head, 266–7, **267**
Melanocytosis, congenital, **82,** 82–3
Melanoma
  choroidal, 268, **268,** 274, **274,** 347–8, **347–8**
  ciliary body *see* Ciliary body melanoma
  conjunctival, 84, **84,** 85, **85**
  iris, 155, **155,** 172–3, **172–3**
  nodular, 34, **34**
  superficial spreading, 37, **37**
  tapioca, 176, **176**
Melanosis
  benign epithelial (racial), 82, **82**
  primary acquired, 82, **82**
Membranes in conjunctivitis, 64, **64**
Meningioma
  optic nerve sheath, 54, **54**
  sphenoidal ridge, 56, **56**
Meretoja syndrome, 130–1, **131,** 391, **391**
Merkel cell (neuroendocrine) carcinoma, 34–5, **35**
Metabolic deposits in stroma and Descemet membrane, 127–30, **127–30**
Metaplastic lashes, 5, **5**
Metastases
  choroidal carcinoma, 349, **349**
  Ewing sarcoma, 50
  iris carcinoma, 173, **173**
  multiple iris, 176, **176**
  neuroblastoma, 50, **50**
  proptosis, 53
  skull lytic bony lesions, 404, **404**
  Wilms tumour, 50
Methanol toxicity, 263
Methoxyflurane, 323, **323**
Microangiopathy, HIV, 281, **281**
Microbial corneal ulcers, 121–2, **121–2**
Microblepharon, 37, **37**
Microcoria, congenital, 205, **205**
Microcornea, 103, **103**
Microcystoid retinal degeneration, 327, **327**
Microcysts, epithelial, 112, **112**
Microexotropia, 362
Microphakia, 213, **213**
Microphthalmos, 44, **44,** 103–4, **104**
  cataracts in neonates, 221, **221**
  with cyst, 51, **51**
Microspherophakia, 213, **213**
Microtropia, 358
Mikulicz syndrome, 59
Milia, 27, **27**
Minocycline, 16, **16,** 99
Miotic cysts, 178, **178**
Mixed cell tumour, benign, 58, **58**
Möbius syndrome, 365, **365,** 391, **391**
Molluscum contagiosum
  conjunctival, 69–70, **70**
  eyelids, 27, **27**
Monocular elevator deficit, 365, **365**
Mooren ulcer, 139, **139**
Morning glory anomaly, 254, **254**
Morquio syndrome, 128
Mottling, fundus, 335–6, **335–6**
Mouth, 397–404, **397–404**
  abnormal dentition, 399–400, **399–400**

  abnormal tongue, 402–4, **402–4**
  high-arched palate, 400–2, **400–2**
  ulceration, 397–8, **397–8**
Mucocele, 56, **56,** 60
Mucolipidoses, 111
Mucopolysaccharidoses, 110, **110,** 128, 161
Mucous membrane pemphigoid, 72, **72,** 398, **398**
Mucus fishing syndrome, 78, **78**
Multifocal vitelliform lesions, 344, **344**
Multiple endocrine neoplasia type IIb, 403, **403**
Multiple iris metastases, 176, **176**
Multiple myeloma, 57, 404, **404**
Munson sign, 106, **106**
Muscle overactions, 372–3, **372–3**
Muscle palsies, 370–1, **370–1**
Myasthenia gravis
  facial expression, 389, **389**
  lid ptosis, **20,** 20–1, 25, **25**
  lid retraction, 17, **17**
  muscle palsy, 373–4, **374**
Mydriasis, right
  episodic, 197, **197**
  pharmacological, 196, **196**
Myelination of nerve fibres, 207, **207**
Myogenic and restrictive palsies, acquired, 373–5, **373–5**
Myopia, high, 105, **105,** 253, **253,** 351, **351**
Myopic chorioretinal atrophy, 272, **272**
Myopic macular hole, 326, **326**
Myopic maculopathy, 317, **317**
Myositis, acute, 374
Myotonic dystrophy, 21, **21,** 389, **389**
Myxoedema, 13, **13**

N

Naevus
  acquired, eyelid, 32, **32**
  choroidal, 341, **341**
  congenital, eyelid, 16, **16**
  conjunctival, 83, **83,** 85, **85**
  iris, 172, **172,** 175, **175**
Naevus flammeus, 388, **388**
Naevus of Ota
  eyelid discolouration, 15, **15**
  facial hyperpigmentation, 388, **388**
  heterochromia iridis, 190, **190**
  trabecular hyperpigmentation, 154, **154**
Nail-fold infarcts, 411, **411**
Nails
  dystrophy, 410, **410**
  onycholysis, 409, **409**
  pitting, 409, **409**
  spoon-shaped (koilonychia), 410, **410**
  subungual splinter haemorrhages, 411, **411**
  white (leukonychia), 410, **410**
Nanophthalmos, 44, 103, **103**
Nasal bridge, broad, 361, **361**
Nasopharyngeal carcinoma, 57
Near reflex spasm, 204, **204**
Necrobiosis lipoidica, 417, **417**
Necrotising fasciitis, 10–11, **11**
Necrotising retinitis, 309–10, **309–10**
Neonatal conjunctivitis, 71–2, **71–2**
  bacterial, 71
  chemical, 71, **71**
  chlamydial, 71
  gonococcal, 71, **71**
  herpes simplex type 2, 71
Neonates
  causes and associations of cataract in, 221–3, **221–3**
  conjunctivitis *see* Neonatal conjunctivitis
  premature, retinopathy *see* Retinopathy of prematurity
Neovascular glaucoma, 159, **159**
Neovascularisation, **290–2**
  central and peripheral, 290–1, **290–1**
  choroidal, 272, **272**
  peripheral, 291–2, **291–2**
Nerve fibre myelination, 207, **207**
Neuroblastoma, metastatic, 50, **50**
Neurofibroma, 32, **32**
  plexiform, 48, **48**
Neurofibromatosis type 1, 32, **32,** 161, **161,** 418, **418**
Neurofibromatosis type 2, 228, **228**
Neurogenic palsies, 367–71, **367–71**
  see also specific palsies
Neuropathic (Charcot) arthropathy, 413, **413**
Neuroretinitis, 260, **260,** 288, **288**
Neurotrophic keratopathy, 119, **119**
Nodular basal cell carcinoma, 33, **33**
Nodular meibomian gland carcinoma, 34, **34**
Nodular melanoma, 34, **34**
Nodular squamous cell carcinoma, 34, **34**
Non-Hodgkin lymphoma, 88, **88**
Norrie disease, 206, **206**
North Carolina macular dystrophy, 340, **340**
Norum disease, 128, **128**
Nose, saddle-shaped, 395–6, **395–6**
Nuclear cataract, 230, **230**
Nummular keratitis, 116, **116**
Nystagmus, 378–82, **378–82**
  acquired ocular, 379
  convergence-retraction, 382
  downbeat, 382, **382**
  peripheral vestibular, 380, **380**
  physiological, 378, **378**
  primary congenital, 378–9, **379**
  sidebeat, 380, **380**
  spasmus nutans, 381, **381**
  upbeat, 381, **381**

O

Ocular histoplasmosis syndrome, presumed, 273, **273,** 307, **307**
Ocular motility, 355–82
  congenital syndromes, 363–6, **363–6**
  muscle overactions, 372–3, **372–3**
  torsional deviations in primary position, 371
  see also specific disorder
Ocular myopathy, 21–2, **22**
Ocular siderosis, 191, **191**
Oculocerebrorenal syndrome, 161
Oculocutaneous albinism, 394, **394**
Oculodermal melanocytosis *see* Naevus of Ota
Oculomotor nerve *see* Third nerve
Oculosympathetic palsy, 20, **20,** 23, **23**
Oedema
  corneal, 113–14, **113–14**
  epithelial, 112, **112**
  macular, 310–11, **310–11**
Onchocercal chorioretinitis, 352, **352**
Onycholysis, 409, **409**
Opaque nerve fibres, 271, **271**
Operculum, 243, **243**
Ophthalmitis, sympathetic, 306, **306**
Ophthalmoplegia, chronic progressive external, 375, **375**
Optic atrophy, 256–8, **256–8**
  consecutive, 257, **257**
  primary, 256, **256**
  secondary, 257, **257**
Optic disc *see* Optic nerve head
Optic nerve
  glioma, 48, **48**
  hypoplasia, 297, **297**
Optic nerve head, 249–74
  abnormal disc size, 251–5, **251–5**
    large, 253–5, **253–5**
    small, 251–2, **251–2**
  buried drusen, 260–1, **260–1**
  coloboma, 253–4, **254**
  combined coloboma and pit, 255, **255**
  congenital pit, 253, **253,** 312, **312**
  disc swelling and elevation, 258–63, **258–63**
    bilateral, 260–3, **260–3**
    unilateral, 258–60, **258–60**
  dragged disc, 269–70, **269–70**
  hypoplastic disc, 252, **252**
  juxtapapillary lesions, 270–4, **270–4**
  large cup, 255–6, **255–6**
  tilted disc, 251–2, **251–2**
  tumours and granulomas, 266–8, **266–8**
  vascular abnormalities, 264–5, **264–5**
Optic nerve sheath meningioma, 54, **54**
Optic neuropathy
  arteritic anterior ischaemic, 259, **259**
  compressive thyroid, 263
  non-arteritic anterior ischaemic, 259, **259**
Optico-ciliary shunts, 265, **265**
Orbit, 39–60
  adult disease, 52–7, **52–7**
  atrophy of contents, 45, **45**
  bony abnormalities, 41–4, **41–4**
  childhood disease, 46–52, **46–52**
  cicatrising lesions, 45
  cystic lesions, 51–5, **51–5**
  frozen, 374–5, **375**
  inflammatory disease, 12, **12**
  shallow, associated with craniosynostoses, 42–4, **42–4**

  varices, 55, **55,** 60
  see also specific conditions
Orbital cellulitis, 46, **46,** 53
Orbital fat herniation, 12, **12,** 88, **88**
Orbital floor blow-out fracture, 374, **374**
Orbital rim lesions, 59–60, **59–60**
Osteogenesis imperfecta, 99, **99,** 399, **399**
Osteoma, choroidal, 274, **274**
Osteopetrosis, 49
Oxycephaly, 42, **42**

P

Paget disease, 405, **405**
Palate, high-arched, 400–2, **400–2**
Panuveitis, 238, **238**
  multifocal choroiditis with, **306,** 306–7, 321, **321**
Papillae
  acute conjunctivitis, 65, **65**
  chronic conjunctivitis, 67–8, **67–8**
  reaction in conjunctivitis, 63, **63**
Papillitis, 259, **259**
Papilloedema, 261–2, **262**
  acute, 282, **282,** 288, **288**
Papillomatous eyelid lesions, 30, **30**
Papillophlebitis, 258, **258**
Papules, eyelid, 26–8, **26–8**
Paramedian pontine reticular formation lesion, 376, **376**
Parasitic cysts, 178, **178**
Parinaud dorsal midbrain syndrome, 377, **377**
Parinaud oculoglandular syndrome, 70, **70**
Parinaud syndrome, 18
Parkinsonism, 19, **19,** 390, **390**
Parks three step test, 369, **369**
Parry-Romberg syndrome, 189, **189**
Pavingstone retinal degeneration, 328, **328**
Peau d'orange, 336, **336**
Pellucid marginal degeneration, 107, **107**
Pemphigoid
  bullous, 73, **73**
  mucous membrane, 72, **72,** 398, **398**
Pemphigus vulgaris, 74, **74,** 398, **398**
Penetrating vitreous trauma, 239
Periarteritis, **300,** 300–1
Periocular xanthogranuloma, adult-onset, 14, **14**
Peripheral ulcerative keratitis (PUK), 140, **140**
Periphlebitis, 299–300, **299–300**
Persistent anterior fetal vasculature, 162, 206, **206**
Persistent posterior fetal vasculature, 270, **270**
Peters anomaly
  cataracts in neonates, 221, **221**
  perinatal corneal clouding, 110, **110**
Pfeiffer syndrome, 44, **44**
Phacolytic glaucoma, 149, **149**
Phacomorphic glaucoma, 156, 159, **159**
Phenothiazines, 129, **129**
Phimosis, 420, **420**
Phlyctenulosis, 137, **137**
Phthiriasis palpebrarum, 8–9, **9**
Phthisis bulbi, 44, **44**
Pigmentation
  face, 388, **388**
  see also Hyperpigmentation
Pigment cells, vitreous, 240, **240**
Pigment dispersion syndrome, 153, **153,** 160, 183, **183**
Pigmented conjunctival lesions, 82–4, **82–4**
Pigmented paravenous chorioretinal atrophy, 334–5, **335**
Pilar cyst, 29, **29**
Pilomatricoma (pilomatrixoma), 33, **33**
Pitting, lens, 233, **233**
Placoid pigment epitheliopathy, acute posterior multifocal, 308, **308**
Pleomorphic adenoma, 58, **58**
Plexiform neurofibroma, 48, **48**
Pneumocystosis, choroidal, 305–6, **306**
Poliosis, 4, **4**
Polyarteritis nodosa, 97, **97**
Polychondritis, relapsing *see* Relapsing polychondritis
Polydactyly, 408, **408**
Pontine lesions, bilateral, 376
Porphyria cutanea tarda, 394, **394**
Port wine stain, 388, **388**
Posner-Schlossman syndrome, 159, **159,** 167
Posterior blepharitis, 7, **7**
Posterior capsule fibrosis, 231, **231**
Posterior embryotoxon, 135, **135**
Posterior lenticonus, **214,** 214–15
Posterior orbital encephalocele, 52, **52**
Posterior uveitis, 238, **238**
Postoperative endophthalmitis, acute, 114, **114**
Prayer sign, 407, **407**
Preauricular lymphadenopathy, 64, **64**
Precipitates, inflammatory lens, 233, **233**
Pre-papillary loop, 264, **264**
Preproliferative diabetic retinopathy, 280, **280**
  calibre changes, 297, **298**
Preretinal haemorrhage, 277, **277**
Preseptal cellulitis, 46, **46**
Presumed ocular histoplasmosis syndrome, 273, **273,** 307, **307**
Primary acquired melanosis, 82, **82**
Primary oxalosis type 1, 321, **321**
Progressive bifocal chorioretinal atrophy, 351, **351**
Progressive outer retinal necrosis, 310, **310**
Progressive supranuclear palsy, 377
Proliferative retinopathies, 239, **239**
Proliferative vitreoretinopathy, 330, **330**
Proptosis, 45, **45**
  chronic non-congestive axial, 54–5, **54–5**
  chronic non-congestive non-axial, 55–7, **55–7**
  congestive, 52–3, **52–3**
  dynamic, 60
Prostaglandin analogue, unilateral use of a, 191, **191**
Pseudodendrites, 118, **118**
Pseudodendritic ulceration, 118, **118**
Pseudo-esotropia, 361, **361**
Pseudoexfoliation syndrome
  acquired cataract, 225, **225**
  intraocular pressure, 160
  iris atrophy and translucency, 184, **184**
  trabecular hyperpigmentation, 153, **153**
Pseudo-exotropia, 363, **363**
Pseudogerontoxon, 135, **135**
Pseudo-hypopyon, 149–50, **149–50**
Pseudo-lenticular opacities, 233–4
Pseudophakia, lens opacities in, 231–4, **231–4**
Pseudophakic pigment dispersion, 155, 184, **184**
Pseudo-proptosis, 45, **45**
Pseudo-pterygium, 87, **87**
Pseudo-ptosis, 25–6, **25–6**
Pseudo-tumour, orbital, 46–7, **47**
Pseudoxanthoma elasticum
  AD type 2, 100, **100**
  high-arched palate, 400, **400**
Psoriatic arthritis, 169, **169,** 412, **412**
Pterygium, 87, **87**
Ptosis
  brow, 25, **25**
  eyelash, 5, **5,** 26, **26**
  eyelid *see* Eyelid ptosis
Pucker, 320, **320**
Punctate epithelial erosions (PEE), 115, **115**
Punctate epithelial keratitis (PEK), **115,** 115–16
Punctate inner choroidopathy, 307, **307,** 316, **316**
Pupil block, 145
  secondary, 160, **160**
Pupillary vascular (Cobb) tufts, 148
Pupils, 193–209
  abnormal reactions, 195–203, **195–203**
  abnormal size, 204–5
    large, 205
    small, 204–5, **204–5**
  right tonic (Adie), 198, **198,** 205
  white, in childhood, 206–9, **206–9**
Purtscher retinopathy, 281, **281**
Pyoderma gangrenosum, 171, **171,** 415, **415**
Pyogenic granuloma, 85–6, **86**

R

Racemose haemangioma, retinal, 296, **296**
Radiation, cataract, 227
Radiation retinopathy, 282, **282,** 286, **286**
Radiation telangiectasia, 81, **81**
Raynaud phenomenon, 408, **408**
Reactive lymphoid hyperplasia, 88, **88**
Reactive pseudo-epitheliomatous hyperplasia, 88, **88**
Reiger syndrome
  abnormal dentition, 399, **399**
  Axenfeld–Rieger anomaly, 182, **182**
Reis-Bückler dystrophy, 126, **126**

Reiter syndrome
  anterior uveitis, 168, **168**
  arthritis, 413, **413**
  conjunctivitis, 74, **74**
  corneal infiltrates, 137
  genitalia ulceration, 419, **419**
  mouth ulceration, 397, **397**
Relapsing polychondritis
  anterior uveitis, 171, **171**
  saddle-shaped nose, 396, **396**
  scleritis, 96–7, **97**
Renal failure, 14, **14**
Rendu-Osler-Weber disease, **81**, 404, **404**
Restrictive palsies, acquired, 373–5, **373–5**
Restrictive thyroid myopathy, 373, **373**
Retention cyst, conjunctival, 89, **89**
Retina
  astrocytoma, 346, **346**
  benign flecked, 339, **339**
  calibre changes, 297–8, **297–8**
  coloboma, 343, **343**
  detachment *see* Retinal detachment
  emboli, 292–3, **292–3**
  focal elevated lesions, 346–7, **346–7**
  hamartoma, 267, **267**, 270, **270**
  hard exudates, 284–7, **284–7**
  inflammation *see* Retinitis
  miscellaneous disorders, 239, **239**
  necrosis *see* Retinal necrosis
  peripheral degenerations, 327–9, **327–9**
    innocuous, 327–8, **327–8**
    predisposing, 328–9, **328–9**
  solitary fragments, 243, **243**
  telangiectasia, 286, **286**
  vascular malformations, 294–5, **294–5**
  vascular tumours, 295–6, **295–6**
Retinal artery
  macroaneurysm, 286, **286**, 294, **294**
  occlusion, calibre changes, 297, **297**
Retinal detachment, 329–32, **329–32**
  differential diagnosis, 331–2, **331–2**
  exudative, 331, **331**
  rhegmatogenous, 329–30, **329–30**
  tractional, 330, **330**
Retinal macrovessel, congenital, 295, **295**
Retinal necrosis
  acute, **309**, 309–10
  progressive outer, 310, **310**
Retinal pigment epithelium (RPE)
  congenital hypertrophy, 271, **271**, 343, **343**
    atypical, 341, **341**
    grouped, 333, **333**
    typical, 340, **340**
  detachment, 313, **313**
  hamartoma, 267, **267**, 270, **270**, 341, **341**
Retinal vein occlusion, 280, **280**
  calibre changes, 297, **298**
  old branch, 285, **285**
Retinitis
  cytomegalovirus, 289, **289**, 309, **309**
  necrotising, 309–10, **309–10**
  serous detachment, 313, **313**
Retinitis pigmentosa, 297, **297**, 334, **334**
Retinitis punctata albescens, 339, **339**
Retinoblastoma, 346, **346**
  buphthalmos, 162, **162**
  endophytic, 240, **240**
  iris invasion, 176, **176**
  neglected, 50
  pseudo-hypopyon, 149, **149**
  white pupil in childhood, 208, **208**
Retinochoroiditis, old, 342, **342**
Retinochoroidopathy, birdshot, 308, **308**
Retinoma, 346–7, **347**
Retinopathies
  AD crystalline, 322, **322**
  diabetic *see* Diabetic retinopathy
  idiopathic central serous, 312, **312**
  preproliferative diabetic, 280, **280**
  proliferative, 239, **239**
  Purtscher, 281, **281**
  radiation, 282, **282**, 286, **286**
  rubella, 335, **335**
  severe hypertensive, 281, **281**
  sickle cell, 292, **292**
  siderosis, 352, **352**
Retinopathy of prematurity, 269, **269**
  calibre changes, 297, **298**
  cicatrical, 208, **208**
  neovascularisation, 292, **292**
Retinoschisis, degenerative, 331, **331**
Retro-dots, lenticular, 230, **230**
Rhabdomyosarcoma, 13, **13**, 49, **49**
Rhegmatogenous retinal detachment, 329–30, **329–30**
Rheumatoid arthritis, 412, **412**
  corneal thinning and ulceration, 140, **140**
  scleritis, 96, **96**
Rodent ulcer, 35, **35**
Rosacea keratitis, 136, **136**
Rosai-Dorfman disease, 14, **14**
Roth spots, 278, **278**
Rubella
  intrauterine, 109
  retinopathy, 335, **335**
Rubella iritis, 185, **185**
Rubeosis iridis, 180, **180**
  ectropion uveae, 179, **179**
  hyphaema, 148, **148**
  neovascular glaucoma, 159, **159**
Rubeotic angle closure, 157, **157**
Rust ring, 123

S

Saddle-shaped nose, 395–6, **395–6**
Salzmann nodular degeneration, 125, **125**
Sampaolesi line, 153
Sarcoid granuloma, 268, **268**
Sarcoidosis
  acute, 170, **170**
  chorioretinitis, 302–3, **302–3**
  chronic, 170, **170**
  facial signs, 386, **386**
  lacrimal gland enlargement, 59
  neovascularisation, 292
Scheie syndrome, 128
Sclera/episclera, 91–100
  anterior necrotising disease, 95, **95**
  anterior non-necrotising disease, 93–4, **93–4**
  blue, 99–100, **99–100**
  discolouration, 98–100, **98–100**
  inflammation *see* Episcleritis; Scleritis
Scleral hyaline plaque, 98, **98**
Scleritis
  causes and systemic associations of, 96–8, **96–8**
  diffuse, 93, **93**
  necrotising, with inflammation, 95, **95**
  nodular, 94, **94**
  posterior, 95, **95**
Sclerocornea, **109**, 109–10
Sclerodactyly, 408, **408**
Scleroderma, 76, **76**, 283, **283**
Scleromalacia perforans, 95, **95**
Sclerosing basal cell carcinoma, 36, **36**
Sclerosing keratitis, 137–8, **138**
S-cone syndrome, enhanced, 335, **335**
Sebaceous cyst, 29, **29**
Seborrhoeic blepharitis, 7, **7**
Seborrhoeic keratosis, 30, **30**
Seidel test, 144, **144**
Senile keratosis, 32, **32**
Senile verruca, 30, **30**
Sentinel vessels, 79, **79**
Serous detachment, 312–13, **312–13**
Serpiginous chorioretinopathy, 352, **352**
Serpiginous choroidopathy, 274, **274**, 309, **309**
Setting sun sign, 18, **18**
Sickle cell retinopathy, 292, **292**
Siderosis, ocular, 191, **191**
Siderosis retinopathy, 352, **352**
Silicone oil, emulsified, 150, **150**
Sinus histiocytosis, 49
Sinus tumour, 50, 56, **56**
Sipple syndrome, 403, **403**
Sixth nerve palsy, 369–70, **369–70**
Sjögren-Larson syndrome, 322, **322**
Sjögren syndrome, 59, 402, **402**
Skin, 413–18, **413–18**
  multiple tumours, 418, **418**
  thin hyperelastic, 416, **416**
  see also specific disorder
Skull, 404–5, **404–5**
  large, 405, **405**
  lytic bony lesions, 404–5, **404–5**
Snailtrack retinal degeneration, 328, **328**
Snowflake cataract, 228, **228**
Soemmering ring, 232, **232**
Soft drusen, 337, **337**
Solar keratosis, 32, **32**
Sorsby pseudo-inflammatory dystrophy, 340, **340**
Spasmus nutans, 381, **381**
Sphenoidal ridge meningioma, 56, **56**

Spheroidal degeneration, 124–5, **125**
Sphingolipidoses, 345, **345**
Spreading meibomian gland carcinoma, 37, **37**
Squamous cell carcinoma
  invasive conjunctival, 87, **87**
  nodular, 34, **34**
  ulcerating, 35, **35**
Squamous cell papilloma
  conjunctival, 85, **85**
  eyelid, 30, **30**
Staphylococcal blepharitis, 7, **7**
Staphyloma, congenital anterior, 110, **110**
Stargardt macular dystrophy, 317–18, **318,** 319
Steele-Richardson-Olszewski syndrome, 377
Stellate cataract, 229, **229**
Stellate maculopathy, 287–9, **287–9**
Stevens-Johnson syndrome
  conjunctivitis, 73, **73,** 74
  genitalia ulceration, 419, **419**
  mouth ulceration, 397, **397**
Stickler syndrome
  ectopia lentis, 217
  empty vitreous, 246, **246**
  high-arched palate, 402, **402**
Stocker line, 123
Strabismus fixus, 366, **366**
Stroma
  anterior
    degenerations in, 124–6, **124–6**
    dystrophies, 126–7, **126–7**
  cystic lesions, 177, **177**
  deposits in, 127–30, **127–30**
  dystrophies, 111, 130–2, **130–2**
Stromal keratitis, acute, 138, **138**
Sturge-Weber syndrome
  childhood glaucoma, 161, **161**
  conjunctiva vascular dilatation and malformation, 80, **80**
  eyelid discolouration, 15, **15**
  heterochromia iridis, 190, **190**
Stye, 31, **31**
Subacute angle closure, 155
Subhyaloid haemorrhage, 277, **277**
Subperiosteal abscess, 60
Subperiosteal haematoma, 59
Subretinal fibrosis, 320, **320**
  diffuse, 321, **321**
Subretinal haemorrhages, 278–9, **279**
Sub-RPE haemorrhages, 279, **279**
Subungual splinter haemorrhages, 411, **411**
Sulfite oxidase deficiency, 217
Summerskill sign, 19
Superficial dermoid cyst, 29, **29,** 59, **59**
Superficial punctate keratitis (SPK), 115, **115**
Superficial spreading melanoma, 37, **37**
Superior limbic keratoconjunctivitis, 68, **68,** 117, **117**
Superior rectus palsy, 370, **370**
Surgically-induced scleritis, 96, **96**
Sutural childhood cataract, 220, **220**
Sympathetic ophthalmitis, 306, **306**
Sympathomimetic drops, topical, 19
Synchysis scintillans, 242
Syndactyly, 409, **409**
Syphilis
  acquired
    chorioretinitis, 334, **334**
    genitalia ulceration, 419, **419**
    inflammatory fundus lesions, 305, **305**
    secondary, 398, **398**
  congenital
    abnormal dentition, 399, **399**
    fundus mottling, 335, **335**
    high-arched palate, 401, **401**
    interstitial keratitis, 133, **133**
    saddle-shaped nose, 395, **395**
Syringomas, 27, **27**
Systemic lupus erythematosus
  cotton-wool spots, 283, **283**
  facial signs, 386, **386**
  scleritis, 97–8, **98**
Systemic sclerosis, 390, **390**
Systemic signs, 383–420
  arthritis, 412–13, **412–13**
  face, 385–98, **385–98**
  genitalia, 418–20, **418–20**
  hands, 406–9, **406–9**
  mouth, 397–404, **397–404**
  nails, 409–11, **409–11**
  skin, 413–18, **413–18**
  skull, 404–5, **404–5**

T

Tamoxifen, 323, **323**
Tangier disease, 128
Tapioca melanoma, 176, **176**
Tectal (dorsal midbrain) pupils, 202, **202**
Teeth, abnormal, 399–400, **399–400**
  caries, 399, **399**
  staining and hypoplasia, 400, **400**
Telangiectasia, 239
  hereditary haemorrhagic, 81, **81**
  radiation, 81, **81**
  retinal, 286, **286**
  skin signs, 417, **417**
  2A bilateral idiopathic acquired parafoveolar, 324, **324**
Teratoma, 49
Terrien marginal degeneration, 139, **139**
Terson syndrome, 239
Thiel-Behnke dystrophy, 126, **126**
Thin hyperelastic skin, 416, **416**
Thioridazine retinotoxicity, 333, **333**
Third nerve misdirection, 17, **17,** 20
Third nerve palsy
  ptosis, 19, **19,** 23, **23**
  right, 200, **200**
  signs, **367,** 367–8
Thyroid eye disease
  in children, 47, **47**
  congestive proptosis, 52, **52**
  late, 59
  lid retraction, 17, **17**
  non-congestive proptosis, 54, **54**
Thyroid myopathy, restrictive, 373, **373**
Thyrotoxicosis, 75, **75**
Tilted disc, 251–2, **251–2**
Tobacco dust, 240, **240**
Tongue, abnormal, 402–4, **402–4**
Torsional deviations in primary position, 371
Toxic epidermal necrolysis, 73, **73**
Toxocara granuloma
  focal elevated retinal lesions, 347, **347**
  optic nerve head, 268, **268,** 273, **273**
  peripheral, 269, **269**
Toxocariasis, 302, **302**
Toxoplasmosis, 273, **273,** 301, **301**
Trabecular hyperpigmentation, 153–5, **153–5**
Trabeculodysgenesis, isolated, 160, **160**
Trachoma, 69, **69**
Tractional retinal detachment, 330, **330**
Transient eye popping reflex, 19
Trauma
  birth, 109, **109**
  blunt *see* Blunt trauma
  iatrogenic vitreous, 239
  macular hole, 326, **326**
  penetrating vitreous, 239
  vitreous, 239, **239**
Trichiasis, 4, **4**
Trichilemmoma, 33, **33**
Trichomegaly, 6, **6**
True exfoliation, 225, **225**
Tuberculosis, 305, **305**
Tuberous sclerosis, 387, **387**
Tumour cells, vitreous, 240, **240**
Tumours
  benign orbital, in children, 47–9, **47–9**
  intraocular, 162, **162**
  iris, 148, **148,** 179, **179,** 191, **191**
  juxtapapillary, 274, **274**
  malignant eyelid, 57
  malignant orbital, in children, 49–50, **49–50**
  multiple skin, 418, **418**
  optic nerve head, 266–8, **266–8**
  rapidly growing eyelid, 13, **13**
  retinal vascular, 295–6, **295–6**
  ulcerating eyelid, 35–6, **35–6**
  see also specific tumours
Turner syndrome, 100, **100,** 393, **393**

U

Ulcerating eyelid tumours, 35–6, **35–6**
Ulceration
  corneal *see* Corneal ulceration
  genitalia, 419, **419**
  mouth, 397–8, **397–8**
Ulcerative colitis, 138, 171
Up-shoot in abduction, 372
Uraemia, 19
Urethritis, 418, **418**
Uveal effusion syndrome, 332, **332**
Uveal melanocytic proliferation, bilateral diffuse, 342, **342**

Uveitis
  anterior *see* Anterior uveitis
  intermediate, 237, **237**
  panuveitis, 238, **238**
  posterior, 238, **238**

V

Vacuoles, lenticular, 231, **231**
Varices
  conjunctival, 79, **79**
  orbital, 55, **55,** 60
Vasculitis
  fundus, 299–301, **299–301**
  optic disc, 258
  skin signs, 416, **416**
Vernal keratoconjunctivitis, **67,** 67–8
Vertical gaze palsy, 377, **377**
Vesicles
  corneal, 112, **112**
  eyelid, 26–8, **26–8**
Vesiculobullous dermatoses, 415, **415**
Visual symptoms, differential diagnosis, 354
Vitelliform lesions, multifocal, 344, **344**
Vitelliform macular dystrophy, 344, **344**
Vitiligo, 16, **16,** 414, **414**
Vitreoretinochoroidopathy, autosomal dominant, 245
Vitreoretinopathies
  autosomal dominant neovascular inflammatory, 245
  erosive, 245
  familial exudative, 245, **245**
  hereditary, 245–7, **245–7**
  proliferative, 330, **330**
Vitreous, 235–47
  acute posterior detachment, 238, **238,** 241, **241**
  cells, 237–40, **237–40**
  cyst, 243, **243**
  hereditary vitreoretinopathies, 245–7, **245–7**
  opacities, 241–4, **241–4**
  see also specific disorders
Vogt-Koyanagi-Harada syndrome
  anterior uveitis, 170, **170**
  chorioretinitis, 304–5, **304–5**
  diffuse subretinal fibrosis, 321, **321**
Vogt striae, 106, **106**
Vogt white limbal girdle, 134, **134**
Vortex keratopathy, 123, **123**
V pattern deviation, 366, **366**

W

Waardenburg syndrome, 189, **189**
Wagner disease, 247, **247**
Water clefts, lenticular, 231, **231**
Wegener granulomatosis, 97, **97,** 395, **395**
Weill-Marchesani syndrome, 216, **216,** 406, **406**
Wet age-related macular degeneration, 285, **285,** 289, **289**
White-without-pressure retinal degeneration, 328–9, **329**
Wilms tumour, metastatic, 50

X

Xanthelasma, 28, **28**
Xanthogranuloma
  adult-onset periocular, 14, **14**
  childhood, 162
Xeroderma pigmentosum, 387, **387,** 418, **418**
X pattern deviation, 366

Y

Y pattern deviation, 367